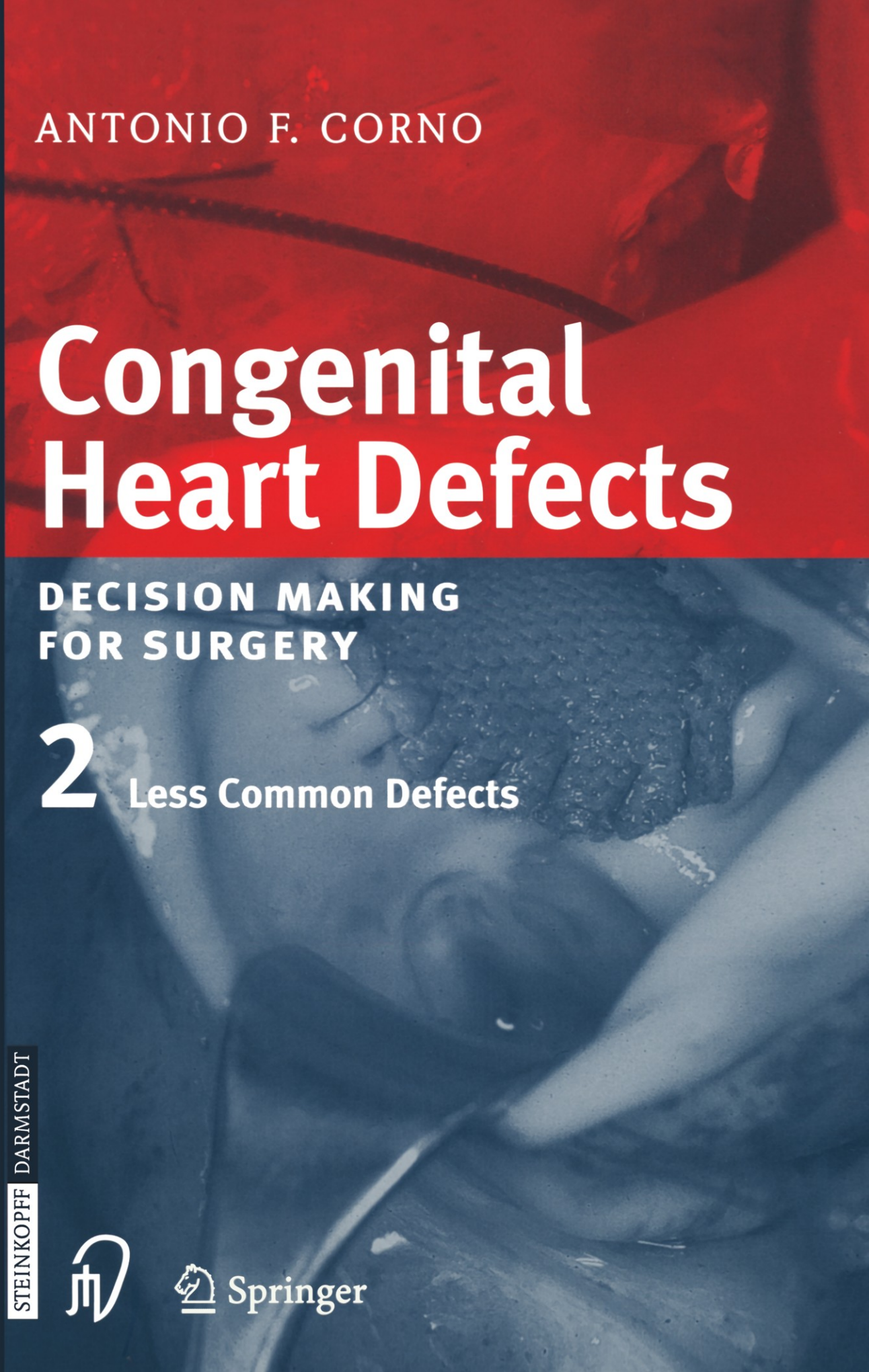
ANTONIO F. CORNO

Congenital Heart Defects

DECISION MAKING FOR SURGERY

2 Less Common Defects



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Antonio F. Corno

Congenital Heart Defects

Decision Making for Cardiac Surgery

Volume 2 Less Common Defects

Dedicated to my loved children
Federica, Laura and Jonathan

Antonio F. Corno

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Decision Making for Cardiac Surgery

Volume 2 Less Common Defects

Foreword by Pedro J. del Nido

With 145 Figures in 340 Separate Illustrations

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Foreword

Nearly four decades have passed since attempts at correcting intracardiac defects in children first began to succeed. In that time great advances have occurred in our field, including improved diagnostic techniques as well as our ability to manage even the most complex heart defects with relatively low risk and an expected outcome that was unimaginable just twenty years ago. New non-invasive techniques for imaging the heart have largely supplanted more invasive methods such as angiography for the diagnosis of most heart defects. Continued advances in this area promise to provide the practitioner accurate quantitative information regarding the hemodynamics and pathophysiology of simple and complex heart defects that is often not obtainable even with cardiac catheterization. The surgeon facing surgical correction of an anatomic defect in a child today needs to be familiar with all these imaging and diagnostic modalities in order to have a complete understanding of that child's pathology prior to planning surgical management.

In turn, decision-making regarding timing of intervention, surgical alternatives, and optimal surgical approach remains a challenge for surgeons and cardiologists alike. Knowledge of anatomy and pathophysiology must be complemented with understanding of the potential complications of the various surgical procedures and the expected outcomes. The former has become more difficult as access to morphologic specimens is rapidly becoming a rare phenomenon, and the latter information is not readily available nor is it easily gleaned from most texts. In today's world, the clinician is expected to make important management decisions not only on the basis of personal experience but also on the accumulated knowledge, often over decades, of rapidly evolving practice.

Although there is a plethora of textbooks available that provide an encyclopedic volume of information regarding all the described congenital heart defects, there is a distinct lack of practical information on how decisions regarding management are made in order to optimize outcome in our patients. The author of this text has taken great care to provide this information in a logical step by step fashion, including interpretation of diagnostic tests, background on the morphologic features and common clinical presentation, as well as operative approaches and techniques along with potential pitfalls. This valuable text is arranged in a sequential manner to cover more common defects in the initial volume, followed by coverage of less common congenital heart defects. The emphasis is on practical information, with ample illustrations which are essential for an understanding of the morphologic features of the defects as well as the theory and practice of surgical repair. The focus on

decision-making is particularly useful for clinicians treating children with congenital heart defects and this text will be of value to surgeons and cardiologists early in their career, as well as those with experience wishing to update their knowledge.

It gives me great pleasure to provide these reflections as an introduction to this valuable text. I commend the author for focusing on the practical issues involved in the management of these many complex defects. Doctor Corno is to be congratulated for writing a volume which will surely become a reference text for clinicians in our field for many years to come.

Children's Hospital – Boston, Massachusetts, USA PEDRO J. DEL NIDO, M.D.
August 2004

Preface

Following the program agreed with the Publisher, one year after the publication of Volume 1 dealing with the most frequent congenital heart defects, Volume 2 appears with the elements for the decision making process in the surgical treatment of the less common heart malformations.

The schema utilized for the construction of the book remain unchanged. The less frequent congenital heart defects are presented with each chapter devoted to a single malformation, with incidence, morphology, associated anomalies, pathophysiology, diagnosis (including clinical pattern, electrocardiogram, chest X-ray, echocardiogram, and cardiac catheterization with angiography), indications for surgical treatment, details of surgical treatment, potential complications and literature references.

In comparison with Volume 1 the technical quality of the reproduction of the schematic drawings has been substantially improved, as well as the selection of echocardiographic and angiographic images. With regard to the diagnostic tools, photographs with magnetic resonance imaging and computerized tomography have been selected to add further information on specific defects. More space has been dedicated to the intra-operative photographs, since these were particularly appreciated in Volume 1. Finally, the number of references has been extensively increased for each chapter, following a request frequently received from the readers.

As for Volume 1, the acknowledgements to those who contributed to my knowledge in the field of congenital heart defects remain valid for Volume 2. In addition, I would like to thank the individuals providing me with help and support for the preparation of this volume: Bruno Marino and Gaetano Thiene once again allowed me to illustrate the details of cardiac morphology by reproducing wonderful illustrations coming either from their "Atlante di anatomia ecocardiografica delle cardiopatie congenite" or from their personal collections. Michael Rigby kindly made available his library of echocardiographic images of the most rare congenital heart defects. Adriano Carotti, who was a junior fellow when I was in Rome, quickly became a talented surgeon, expert in Pulmonary Atresia with Ventricular Septal Defect. He kindly revised the relevant chapter with constructive criticism.

Philippe Monnier and Florian Lang, ENT surgeons of Lausanne who developed the technique of slide tracheoplasty for long-segment tracheal stenosis associated with the simultaneous repair of congenital heart defects, contributed to the chapter "Slings and rings" with their precious input. Philippe Clavel, once again with much patience, prepared the graphics following my continuous requests for improvements.

As for Volume 1, Bruno Marino kindly reviewed the entire text, giving particular care to the morphological and cardiologic components. My current Chairman, Ludwig K. von Segesser, with his usual attention to details, stimulated most of the improvements over Volume 1. He was instrumental in respecting the deadline for completion, by asking with increasing frequency how my job was progressing.

A special acknowledgement is due to the Publishers, particularly the CEO Dr. Thomas Thiekötter, Sabine Ibkendanz (head of medical editing) and Sabine Scheffler (marketing dpt.). A separate mention is reserved for Oliver Frohmeyer (medical editing), who step by step guided me from the beginning through this adventure of the preparation of these books as a single author. Without their continuous support, particularly during the most difficult moments, these books simply would not exist.

During the preparation of this volume, I had the great chance of personally meeting the individual who entirely changed my vision of the cardiac anatomy and physiology: Dr. Francisco Torrent-Guasp. Francisco, now a retired general practitioner in the Spanish village of Denia, not far from Alicante, spent 50 years of his life cultivating his hobby: the investigation of the normal cardiac anatomy and physiology. Without any support from a hospital or university, and despite the nihilism and sometimes the sarcasm of most of the other colleagues, he was able through the dissection of more than 1000 hearts to reveal the existence of the "ventricular myocardial band", the unique myocardial structure constituting right and left ventricles. Despite his initial observations having been published 50 years ago when he was a medical student, only within the last few years have his theories become available to the scientific public. Several researchers have been recently stimulated by his work to further investigate the ventricular myocardial band. Francisco Torrent-Guasp represents to me the ideal prototype of researcher, who just following his intellectual curiosity and his passion for knowledge, independent from the opinion or the support of anyone else, was able to pursue the truth by "beating to death" the topic attracting his attention. Currently, the potential consequences on the management of congenital heart defects based on the observations by Francisco on the normal cardiac anatomy and physiology are only the object of speculation. We are only discovering the tip of the iceberg, but thanks to Francisco the way has been opened up.

Finally, I would like to recognize the unconditional support and indispensable encouragement from my entire family to complete this second book; thank you Josie, Federica, Laura and Jonathan.

Lausanne, Switzerland, August 2004 ANTONIO F. CORNO

Acknowledgments

Since this book is the result of my personal experience, I would like to thank all the individuals who contributed to developing my knowledge in the field of congenital heart defects.

Acknowledgment begins with all the sick children and their families encountered during my professional life.

Then to all the teachers who contributed to my professional training, colleagues, nurses and technicians met during my career: from everybody I have learned something, from most I learned how, when and what to do in the presence of a child with a congenital heart defect; from others what should not be done which is also extremely important.

Morphology: Thanks to the endless enthusiasm of Gaetano Thiene and his huge specimen collection at the University of Padova, where I received the rudiments of the morphology of congenital heart defects; he spent a tremendous amount of time and effort teaching the anatomy of the most frequent cardiac malformations.

While in Amsterdam for my surgical training, Anton E. Becker, another outstanding cardiac pathologist came into the operating room, scrubbed with the surgeons, explained the details of the intra-cardiac morphology and provided precious suggestions for surgical repair.

Impossible not to mention are Robert H. Anderson and Richard van Praagh for their educational books and articles as well workshops with practical demonstrations. Robert H. Anderson supported the production of this book and provided substantial input for the preparation of a few chapters.

Pathophysiology: Understanding the pathophysiology is essential in the process of decision making for congenital heart defects. At the University of California, Los Angeles, Jay M. Jarmakani and particularly William F. Friedman were always available to explain the myocardial function in normal and sick children. Particularly important were the lessons repeatedly learned from Abraham M. Rudolph, with both editions of his remarkable book and numerous outstanding papers.

Clinical pattern: Piero Fancini, in Milan, explained cardiac murmurs, electrocardiograms and chest X-rays, Filippo Casolo taught the basics of angiocardiography. Tom G. Losekoot continued this education in Amsterdam on hemodynamics, followed by Roberta G. Williams at the University of California, Los Angeles, on pre-operative and post-operative echocardiography and

Joseph K. Perloff on the problems of the growing population of adults with congenital heart defects.

A substantial part of my knowledge on clinical problems has been acquired by daily practice, particularly in the pediatric hospital "Bambino Gesú", Rome, which all the pediatric cardiologists contributed to, particularly Bruno Marino and Salvatore Giannico. A similar positive experience occurred years later in Glasgow, with Neil Wilson.

Surgery: The beginning of my training was at the University of Padova, with the late Vincenzo Gallucci, who taught me how to repair an atrioventricular septal defect and a tetralogy of Fallot.

Further important progress was the exposure to daily practice with Carlo Marcelletti, not only in surgery but also in pre-operative evaluation as well as in post-operative care. Most of the intra-operative photographs of this book have been taken during the period spent with him.

Other surgeons substantially contributed to improving my surgical expertise:

- Hillel Laks, at the University of California, Los Angeles, who was very demanding and meticulous.
- Yves Lecompte, in Paris, thanks to his straightforward style, corrected when needed, before actually teaching how surgery should be done and sometimes how to be inventive. His essential observation was that "every patient is unique".
- Ludwig K. von Segesser, my current Chairman, is one of the few remaining surgeons able to operate on a neonate of 2 kg with transposition of the great arteries and single coronary artery, a 86 year-old patient with the rupture of a thoraco-abdominal aneurysm and to support the decision for a Ross operation on a young adult with 4 previous sternotomies. Not to mention his constant intellectual curiosity to develop new techniques and tools in the research laboratory before clinical application. He also pushed me, more than anyone else, to complete this book.

Other surgeons participated in extending my surgical knowledge during meetings and workshops, like all the colleagues of the European Congenital Heart Surgeons Foundation.

Other contributions came from the visits to the hospitals of Aldo R. Castañeda, Mark R. de Leval, Richard A. Jonas, William I. Norwood, Lucio Parenzan, Claude Planché, Jan M. Quaegebeur, Jaroslav Stark and Pascal Vouhé.

Cardiopulmonary bypass: Yves Durandy, in Paris, demonstrated that cardiopulmonary bypass can and should be performed in a way very similar to the physiological conditions.

Post-operative care: I learned from all the colleagues and nurses in the pediatric hospital "Bambino Gesú", Roma.

Yves Durandy proved that the post-operative period should respect closely the physiological conditions with the fewest medications and interventions.

Leonardo Milella, in Glasgow, confirmed that it is possible for the anesthesist and surgeon to collaborate very well in the post-operative care, with evident advantage for the patients.

Research: Several individuals were very important in different periods of my experience with experimental and clinical research, but none as much as Gerald D. Buckberg, University of California, Los Angeles, who played a pivotal role in teaching the methodology of research.

Philippe Clavel, Lausanne, contributed to this book with the graphics and a lot of patience with my requests.

Special thanks to Bruno Marino, a friend before being a pediatric cardiologist in Rome, who very kindly reviewed the text of this book and contributed several illustrations.

A profession constitutes only a part of our life: I deeply acknowledge my family's unconditional support, particularly for my wife Josie's patience for the long hours I spent at nights and week-ends working to complete this book.

Lausanne, Switzerland, August 2004 ANTONIO F. CORNO

Table of Contents

Chapter 2.1 Cor triatriatum

Incidence

Cor triatriatum is the 21st most common congenital heart defect (0.1% of all congenital cardiac malformations), but a higher incidence, up to 0.4%, has been reported in autopsies of patients with congenital heart defects.

Morphology

When used in isolation, the term *cor triatriatum* almost always refers to division of the left atrium. The divided right atrium is called *cor triatriatum dexter* (see the end of this chapter). Several patterns exist in which the left atrial chamber is divided, often in association with anomalous venous connections or other lesions. In the great majority of cases, nonetheless, there is a pattern which can be considered as the "classic" lesion. In this variant, the left atrium is divided by a diaphragm (membrane) into two components: 1) the proximal (superior) left atrial chamber, typically thick-walled, somewhat larger than the distal chamber, above the subdividing diaphragm, receiving the four pulmonary veins; 2) the distal (inferior) left atrial chamber, generally thin-walled, with the opening of the fossa ovalis, the left auricular appendage and the mitral valve. The diaphragm between the two components, which may have one or more variably sized openings in it, is usually rather thick and fibromuscular.

Typically, the foramen ovale (which may be deficient, probe-patent or intact) is in actual or potential communication with the distal chamber, and the left auricular appendage is located in the distal chamber; these two features provide a means to distinguish between cor triatriatum and supravalvular mitral ring (see chapter "Mitral valve disease"). The severity of the lesion depends upon the size of the orifice between the divided components of the left atrium.

Associated anomalies

Cor triatriatum is seen most frequently as an isolated lesion but it can coexist with any other defect. Notable associations are with persistent left superior vena cava and unroofed coronary sinus. Other associated lesions can be supravalvular mitral ring, atrial septal defect, partial or total anomalous pulmonary venous connection, stenosis of the pulmonary veins, atrioventricular septal defect, mitral valve regurgitation, ventricular septal defect, tetralogy of Fallot, double outlet right ventricle, double discordance, hypoplastic left heart syndrome, aortic valve stenosis, pulmonary stenosis, anomalous origin of the right pulmonary artery from the aorta, aortic coarctation; rarely asplenia or polysplenia.

Pathophysiology

Because of the presence of the fibromuscular diaphragm within the left atrium, the blood flow from the pulmonary veins towards the mitral valve is impeded. Depending on the effective number and size of the opening(s) in the diaphragm dividing the two components

A. F. Corno, *Congenital Heart Defects*

of the left atrium (restrictive or unrestrictive) and on the presence of associated anomalies, the following classifications can be made:

- *unrestrictive opening:* normal hemodynamics or mild pulmonary venous hypertension.
- *restrictive opening:* severe pulmonary venous hypertension, pulmonary arterial hypertension sometimes reaching or exceeding systemic levels.

In the presence of a partially anomalous pulmonary venous connection and/or of a communication between the proximal (superior) left atrial chamber and the right atrium, there is left-to-right shunt at the atrial level, with volume overload of the right heart, and increased pulmonary blood flow.

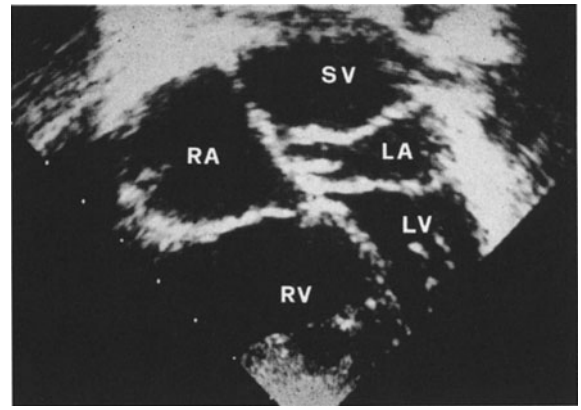

Fig. 2.1.1. Cor triatriatum: echocardiography. The 4-chamber view shows the diaphragm dividing the left atrium into two components: the proximal (superior) left atrial chamber, above the subdividing diaphragm, receiving the four pulmonary veins (*SV* sinus venosus) and the distal (inferior) left atrial chamber with the left auricular appendage and the mitral valve (*LA* distal (inferior) left atrium, *LV* left ventricle, *RA* right atrium, *RV* right ventricle, *SV* sinus venosus (collecting the four pulmonary veins)) (photograph courtesy of Dr. Nicole Sekarski)

Diagnosis

Clinical pattern:

- unrestrictive opening: usually asymptomatic;
- restrictive opening: presentation early in the neonatal period with evidence of low cardiac output syndrome, pulmonary edema, with pallor, poor peripheral pulses, tachypnea, dyspnea, poor feeding, growth failure;
- in the presence of associated left-to-right interatrial shunt: congestive heart failure and/or recurrent upper respiratory infections;
- on auscultation: increased pulmonary component of the second cardiac sound.

Electrocardiogram: right axis deviation, right atrial enlargement, right ventricular hypertrophy.

Chest X-ray: pulmonary venous congestion, cardiomegaly because of right ventricular enlargement.

Echocardiogram: curved diaphragm lying across the left atrium, dividing it into a proximal component with the four pulmo-

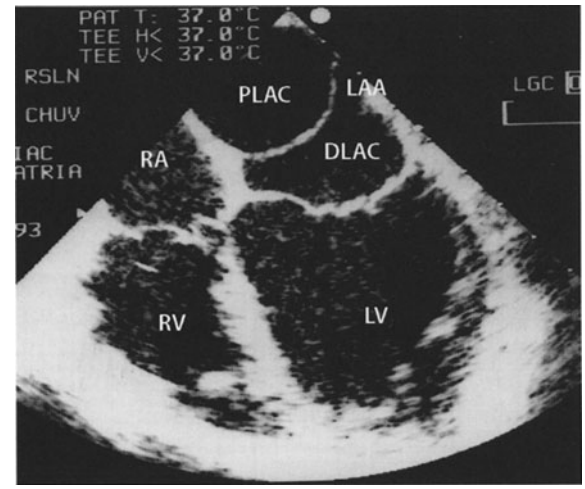

Fig. 2.1.2. Cor triatriatum: echocardiography. Transesophageal echocardiography showing the membrane dividing the left atrium into two chambers: the proximal chamber (superior and posterior) collecting the pulmonary veins and the distal chamber (inferior and anterior) with the left auricular appendage and the mitral valve (*DLAC* distal left atrial chamber, *LAA* left auricular appendage, *LV* left ventricle, *PLAC* proximal left atrial chamber, *RA* right atrium, *RV* right ventricle) (photograph courtesy of Dr. Pierre-Guy Chassot)

nary veins and a distal component with the fossa ovalis and the left auricular appendage (Fig. 2.1.1); transesophageal echocardiography may improve the diagnostic accuracy (Fig. 2.1.2); the diagnosis, easy in the presence of an isolated lesion, may be very difficult in patients with associated cardiac malformations.

Cardiac catheterization: it confirms the presence of a marked increase in pulmonary capillary wedge and pulmonary artery pressure; no longer needed to establish the diagnosis, it may be useful in the presence of associated anomalies and/or to quantitate the pressure gradient between the two left atrial components; in older patients may rule out the presence of pulmonary vascular obstructive disease.

Indications for surgical treatment

Despite the rare discovery of cor triatriatum in adults, in the vast majority of patients the communication between the divided left atrial chambers is severely restrictive, with about 75% of non-treated patients with this defect dying in infancy accordingly with the natural history.

Therefore, in symptomatic neonates the presence of a restrictive opening is an urgent indication for operation, while in symptomatic infants or children there is indication for surgery at the time of diagnosis. In adults with previously unrecognized diagnosis there is indication for surgery only in the presence of symptoms. In older patients the presence of pulmonary vascular obstructive disease must be ruled out.

Surgical treatment (on cardiopulmonary bypass)

Complete resection of the diaphragm, taking care not to injure the mitral valve or the interatrial septum, with approach from the left atrium or from the right atrium (through an already present or a surgically created interatrial communication), depending on the size of the proximal left atrial chamber and on the presence of associated anomalies.

In the classical form of cor triatriatum, the surgical approach through a right atriotomy is recommended, with enlargement of the patent foramen ovale (or interatrial septal defect) to obtain a better exposure of the left atrial diaphragm. After identification and complete resection of the diaphragm, the remaining interatrial communication is closed with an autologous (or heterologous) pericardial patch.

In the left atrial approach, the common pulmonary venous proximal chamber is opened through a vertical incision anterior to the pulmonary veins, and the diaphragm is exposed by appropriate retraction; one or two radial incisions from the opening of the diaphragm outward to the atrial wall or interatrial septum enhance substantially the exposure; the diaphragm is excised only after precise identification of the pulmonary veins.

Potential complications

Inadequate membrane resection, residual atrial septal defect, mitral valve damage, air embolism, supraventricular arrhythmias; in neonates and infants postoperative crises of pulmonary hypertension requiring treatment are frequent. Pulmonary vein stenosis or restenosis at the orifice between the proximal and distal left atrial chambers, generally due to incomplete resection, are quite rare.

Cor triatriatum dexter (divided right atrium)

Cor triatriatum dexter is a term used to describe the partially divided right atrium. It is an extremely rare congenital cardiac malformation and is rarely diagnosed during life unless associated with obstruction of the

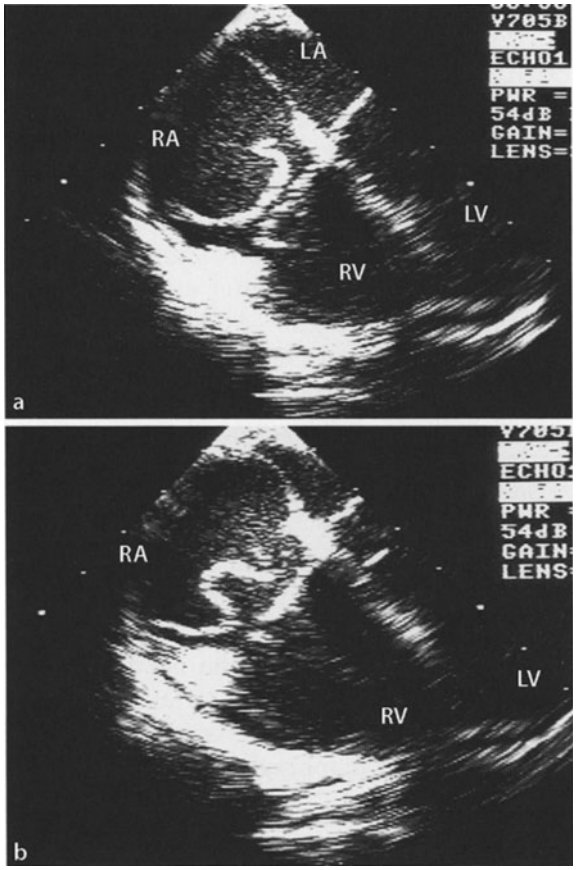

Fig. 2.1.3. Cor triatriatum dexter (=divided right atrium): echocardiography. **a** Pre-operative echocardiography showing the large Eustachian valve in the right atrium during systole, in proximity of the tricuspid valve (*LA* left atrium, *LV* left ventricle, *RA* right atrium, *RV* right ventricle) **b** pre-operative echocardiography in the same patient showing the large Eustachian valve in the right atrium during diastole, in the proximity of the tricuspid valve (reproduced with permission from Corno AF, Bron C, von Segesser LK (1999) Divided right atrium. Diagnosis by echocardiography, and considerations on the functional role of the Eustachian valve. Cardiol Young 9:427–429)

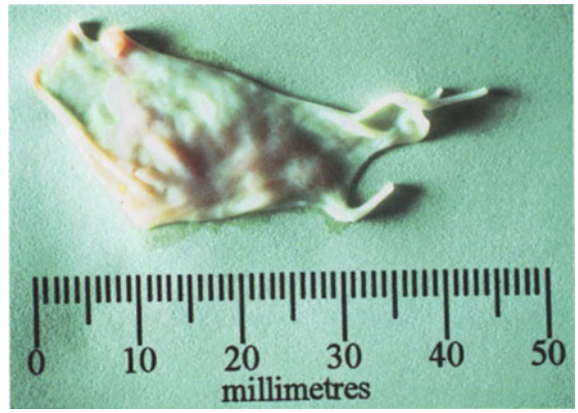

Fig. 2.1.4. Cor triatriatum dexter (=divided right atrium): surgery. Intraoperative photograph of the same patient of Fig. 2.1.3 showing the large abnormal Eustachian valve surgically resected from the right atrium (reproduced with permission from Corno AF, Bron C, von Segesser LK (1999) Divided right atrium. Diagnosis by echocardiography, and considerations on the functional role of the Eustachian valve. Cardiol Young 9:427–429)

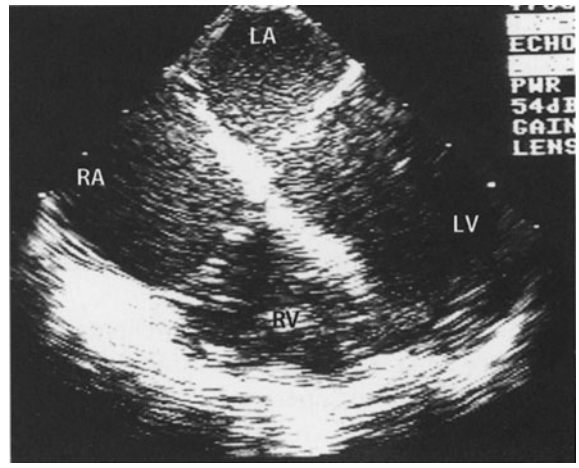

Fig. 2.1.5. Cor triatriatum dexter (=divided right atrium): echocardiography. Postoperative echocardiography of the same patient as in Figs. 2.1.3 and 2.1.4 showing the complete resection of the Eustachian valve from the right atrium (*LA* left atrium *LV* left ventricle, *RA* right atrium, *RV* right ventricle) (reproduced with permission from Corno AF, Bron C, von Segesser LK (1999) Divided right atrium. Diagnosis by echocardiography, and considerations on the functional role of the Eustachian valve. Cardiol Young 9:427–429)

usual pathway of blood to the right ventricle, or with other anomalies on the right heart.

Morphologically the division of the right atrium is due to the persistent right valve of the sinus venosus or Eustachian valve; this valve can persist in part or more extensively. A large Eustachian valve can obstruct the blood flow from the inferior vena cava to the tricuspid valve, reducing the right ventricular filling. In the presence of severe obstruction to the right ventricular filling, particularly in association with an atrial septal defect, the patient can present with cyanosis and poor development of the right ventricle. Rarely the only clinical sign is a supraventricular arrhythmia, or hepatomegaly. Echocar-

diography (Fig. 2.1.3) is the gold standard diagnostic procedure. Surgery is indicated in symptomatic patients, and consists in the simple resection of the prominent Eustachian valve on cardiopulmonary bypass, through a right atriotomy (Fig. 2.1.4). In the absence of major associated congenital lesions, surgery can be contemplated with good long-term prognosis (Fig. 2.1.5).

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Chapter 2.2 Tricuspid atresia

Incidence

Tricuspid atresia is the 19th most common congenital heart defect (0.7–1.6% of all congenital heart defects in clinical series, and up to 2.9% in autopsy series), but is the third most common cyanotic malformation presenting in the neonatal period, after transposition of the great arteries and tetralogy of Fallot. A slight male preponderance exists for tricuspid atresia, particularly in the presence of associated ventriculoarterial discordance.

Morphology (Fig. 2.2.1)

Tricuspid atresia is characterized by the complete absence of a direct communication between the right atrium and the right ventricle (=absent right atrioventricular connection). Tricuspid atresia may range from an imperforate membrane (rarely) to the total absence of the valve, with the area replaced by muscular tissue. The floor of the right atrium is completely muscular, frequently with a tiny dimple (=localized fibrous thickening) in the middle, and is totally separated from the ventricular mass by the atrioventricular sulcus (=absent of any potential right atrioventricular connection). The right atrium is generally dilated, and its wall thickened, particularly in the rare (less than 5% of cases) presence of restrictive interatrial communication (generally the interatrial communication is unrestrictive). The left atrium and the mitral valve are both dilated, since they receive both the pulmonary and the systemic venous returns. The right ventricle is generally poorly developed (sometimes so small that its detection is difficult), and is characterized by total absence of the inlet portion and varying degrees of underdevelopment of the trabecular and infundibular portions. A ventricular septal defect, most frequently of muscular type, is generally present between the hypoplastic right ventricle and the left ventricle, providing access to the rudimentary right ventricle and the pulmonary artery. The atrial situs is almost invariably solitus, and the coronary arteries are generally normal.

The classification of the various forms of tricuspid atresia is based on the type of ventriculoarterial connection and on the amount of antegrade pulmonary blood flow.

Type of ventriculoarterial connection:

- *type I:* normally related great arteries (=ventriculoarterial concordance) (2/3 of infants).
- *type II:* transposition of the great arteries (=ventriculoarterial discordance) (1/3 of infants) (Fig. 2.2.2).

Amount of antegrade pulmonary blood flow:

- *type A:* absence or severe reduction of antegrade pulmonary blood flow, because of pulmonary atresia or stenosis with absent ventricular septal defect (18% of infants); the pulmonary circulation can be totally ductus-dependent.
- *type B:* balanced antegrade pulmonary blood flow (52% of infants), resulting from a moderate degree of obstruction at the level of the ventricular septal defect,

A. F. Corno, *Congenital Heart Defects*

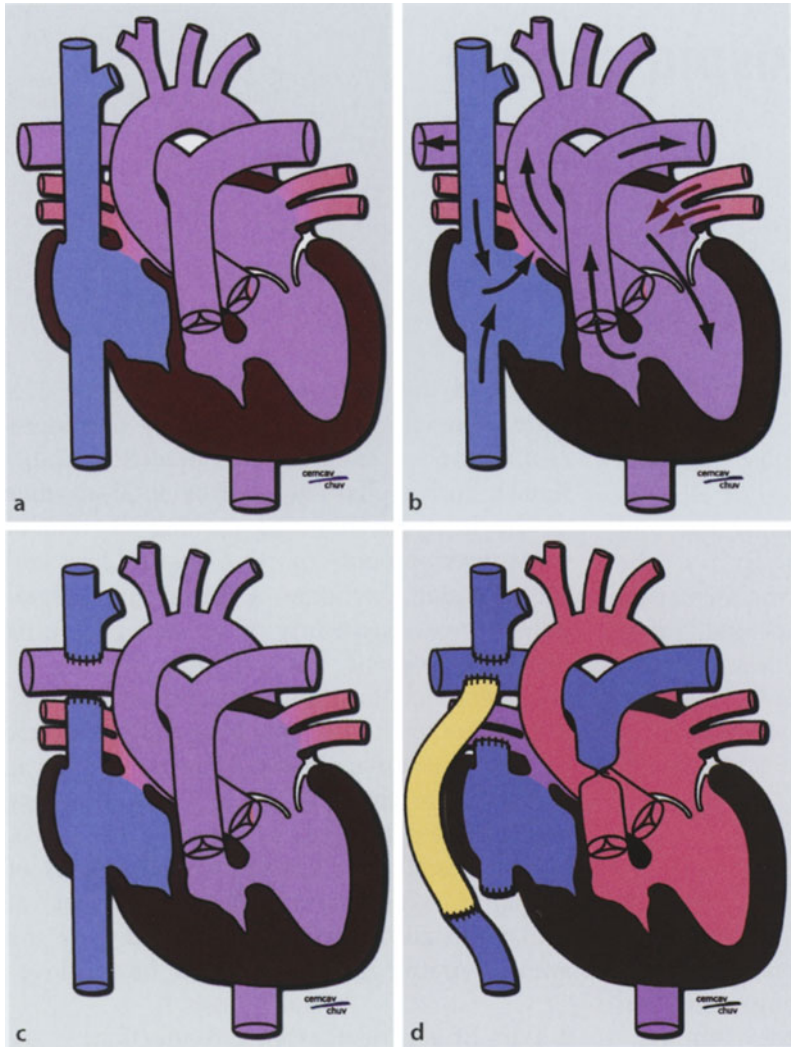


Fig. 2.2.1. Tricuspid atresia: **a** morphology, **b** pathophysiology, and surgery: **c** superior vena cava to right pulmonary artery anastomosis, **d** extra-cardiac total cavopulmonary connection

the right ventricular outflow tract and/or the pulmonary valve, bicuspid in 20% of patients.

- *type C:* unrestricted antegrade pulmonary blood flow (30% of infants), resulting from absence or minimal degree of obstruction at the level of the ventricular septal defect, the right ventricular outflow tract and/or the pulmonary valve.

Associated anomalies

Systemic and pulmonary venous connections are usually normal, with the exception of a persistent left superior vena cava, present in 15% of patients, and partially unroofed coronary sinus with communication between the coronary sinus and the left atrium (1–5% of cases). An atrial septal defect or stretched patent foramen ovale is generally present (the presence of interatrial communication is necessary for survival) and a ventricular septal defect is very frequently present. Ventriculoarterial discordance is present in 1/3 of the patients. Other associated cardiac anomalies are pulmonary stenosis, pulmonary atresia, patent ductus arteriosus), juxtaposition of the auricular appendages (present in 10% of patients with ventriculoarterial discordance), dextrocardia, right aortic arch, aortic coarctation (very rare in

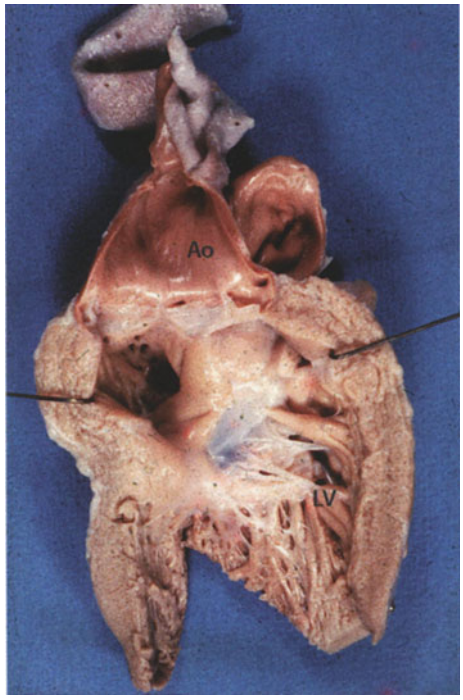


Fig. 2.2.2. Tricuspid atresia: morphology. Morphology of a heart with tricuspid atresia, single ventricular chamber of left ventricular type and ventriculo-arterial discordance (*Ao* aorta, *LV* left ventricle) (photograph courtesy of Dr. Bruno Marino)

patients with ventriculoarterial concordance, but present in up to 30% of patients with ventriculoarterial discordance and restrictive ventricular septal defect).

In 5% of patients there is a very large prominent Eustachian valve, partitioning the right atrium, like in cor triatriatum dexter (see chapter "Cor triatriatum"). There are anedoctical reports of tricuspid atresia with anomalous systemic or pulmonary venous connections, aortic atresia and truncus arteriosus. Situs inversus with ventricular L-loop (mirror imaging pattern) is exceptional. The mitral valve, generally normal, may have a double orifice, isolated anterior cleft or straddling and overriding (see chapter "Straddling atrioventricular valve"). Extracardiac anomalies are present in 13% of children with tricuspid atresia.

Pathophysiology

In patients with tricuspid atresia the left ventricle supports the systemic circulation, either directly (in the presence of normally related great arteries = ventriculoarterial concordance) or indirectly through a ventricular septal defect and the right ventricular outflow tract (in the presence of transposed great arteries = ventriculoarterial discordance).

There is an obligatory right-to-left shunt across the atrial septal defect or stretched patent foramen ovale (always present), with complete mixing of systemic and pulmonary venous return at the left atrial level. Therefore, the first consequence is systemic arterial desaturation, present in all patients with tricuspid atresia, because of the obligatory mixing of the systemic, coronary and pulmonary venous returns in the left atrium.

Restrictive atrial septal defect is more common in the presence of transposed great arteries (= ventriculoarterial discordance). Clinical consequences of flow restriction at the level of the atrial septal defect are low cardiac output, acidosis and severe cyanosis.

The right ventricle is hypoplastic and receives blood from the left ventricle through the ventricular septal defect. The left-sided heart structures (left atrium, mitral valve and left ventricle) are dilated as a consequence of the volume overload due to the combination of systemic and pulmonary venous return. Despite the pathophysiologic enlargement, the left ventricular function generally remains adequate in the early period of the natural history of tricuspid atresia.

Because of the parallel arrangement of the systemic and pulmonary circulations, the pathophysiology of tricuspid atresia is similar to the other heart defects with single-ventricle physiology (= functionally univentricular heart). The flow to each vascular bed is dependent upon their respective resistance. In tricuspid atresia with normally related great arteries (= ventriculoarterial concordance), the antegrade pulmonary blood flow provided by the left ventricle must tra-

verse the ventricular septal defect, the right ventricular outflow tract and the pulmonary valve: each of these structures (or the combination of them) may be responsible for reducing the pulmonary blood flow. The degree of obstruction to the pulmonary blood flow varies from none to complete, with most of neonates having an intermediate degree. In the presence of severe (or complete) obstruction to the antegrade pulmonary blood flow, the pulmonary circulation is totally ductus dependent.

The nature and degree of the obstruction to the pulmonary blood flow may be dynamic and may change over time. Variations in pulmonary vascular resistance and progression of the obstruction at the level of the ventricular septal defect, the right ventricular outflow tract, the pulmonary valve and the patent ductus arteriosus frequently occur within the first few weeks of life.

The most critical situations are with the infant's circulatory balance at one of the two extremes: either systemic desaturation with severe cyanosis or pulmonary overcirculation with congestive heart failure. The balanced pathophysiologic pattern occurs with a QP/QS (pulmonary-to-systemic blood flow ratio) between 1.5 and 2.0, resulting in adequate systemic oxygenation. Lower QP/QS is associated with moderate to severe cyanosis, and higher QP/QS with excessive left ventricular volume overload and congestive heart failure.

A certain degree of left ventricular volume overloading is present in all patients with tricuspid atresia, since the left ventricle is ejecting the entire systemic, coronary and pulmonary outputs.

In the presence of ventriculoarterial discordance there is the potential for either subaortic obstruction or pulmonary outflow tract obstruction, or occasionally the combination of both. With ventriculoarterial discordance, while the subaortic obstruction is generally due to the presence of a restrictive ventricular septal defect (and rarely to the muscular obstruction within the underdeveloped right ventricular outflow tract) and the systemic obstruction to aortic coarctation (rarely to aortic arch interruption) with or without aortic arch hypoplasia, the obstruction to the pulmonary blood flow is mostly due to an obstruction at the level of the hypoplastic right ventricular infundibulum, usually with unrestrictive ventricular septal defect.

The early natural history of tricuspid atresia generally depends upon the degree of obstruction to the pulmonary blood flow.

Diagnosis

Clinical pattern: the clinical pattern depends upon the type of ventriculoarterial connection and the presence and degree of obstruction to the pulmonary blood flow;

- the most common feature is cyanosis, frequently progressive, occurring in the first few weeks or months of life, sometimes with hypoxic spells; severe cyanosis can be present shortly after birth, in the neonates with ductus-dependent pulmonary blood flow;
- neonates may rarely present with low cardiac output, poor peripheral pulses, fast breathing, gray color, prominent neck venous pulsations and hepatomegaly, mostly because of restrictive interatrial communication and/or systemic obstruction at the level of the aortic arch;
- a smaller proportion of infants present at 2-4 months of age with minimal cyanosis but with signs and symptoms of heart failure: dyspnea, tachypnea, tachycardia, fatigue, difficulty in feeding, poor weight gain and perspiration;
- very frequent is the finding of a loud, harsh systolic murmur from the ventricular septal defect or the right ventricular outflow tract; in the presence of associated pulmonary atresia, first and second sounds are combined to a single sound.

Electrocardiogram: very important left axis deviation (with the frontal QRS axis usually from 0° to −90° in the frontal plane), left ventricular hypertrophy (increase in the am-

plitude of S waves in leads V1 and V2) and right atrial enlargement (tall and peaked P waves); normal QRS axis, without left axis deviation, is present in 50% of patients with ventriculoarterial discordance.

Chest X-ray: it is not diagnostic; the cardiac size and the pulmonary vascular markings depend upon the pathophysiologic pattern, with only the right atrium generally dilated, independent of the pathophysiology.

Echocardiogram: it allows definitive diagnosis (Fig. 2.2.3); cross-sectional and Doppler investigations in apical and subcostal 4-chamber views allows the recognition of the absent right atrioventricular connection, the presence and size of the atrial and ventricular septal defects, the type of ventriculoarterial connection and the presence of obstruction to the pulmonary or systemic blood flow.

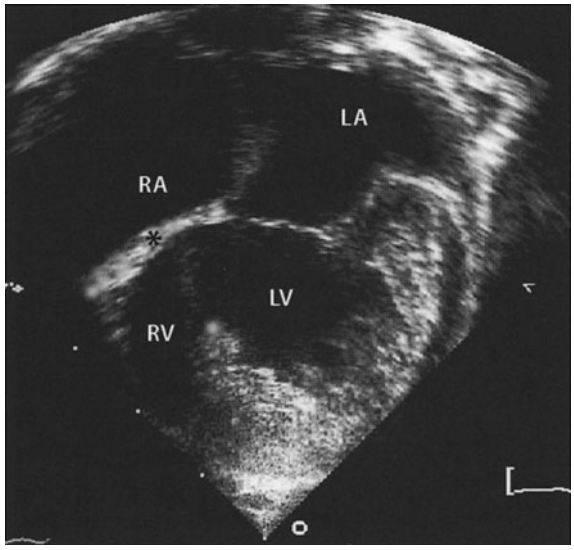


Fig. 2.2.3. Tricuspid atresia: echocardiography. The 4-chamber view showing the absence of the right atrioventricular connection; the muscular tissue of the right atrioventricular sulcus (white star) separates the right atrium from the right ventricle. (*LA* left atrium, *LV* left ventricle, *RA* right atrium, *RV* right ventricle) (photograph courtesy of Dr. Michael Rigby)

Cardiac catheterization: in the neonatal period is indicated in the presence of restrictive atrial septal defect, in order to perform a balloon atrioseptectomy (= Rashkind procedure), in the presence of discrepancies between the echocardiographic diagnosis and the clinical pattern, or with insufficient data from the non-invasive investigations (Fig. 2.2.4); later it is performed to evaluate the pulmonary vascular resistance in view of a cavopulmonary connection, or for percutaneous management of localized narrowing of the branches of the pulmonary arteries.

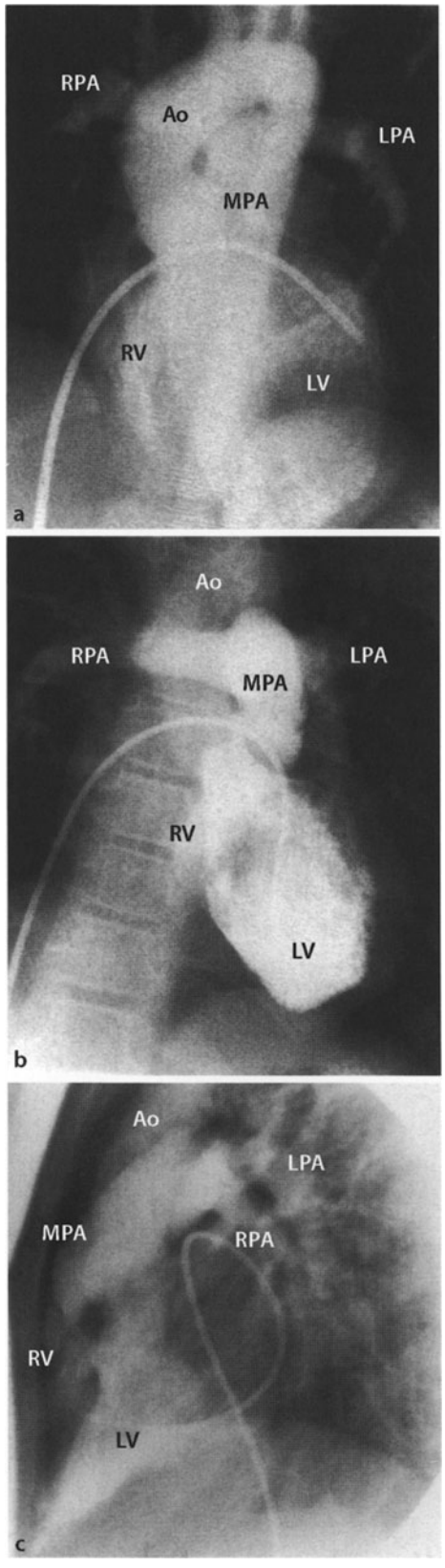
Indications for surgical treatment

Without treatment, patients with tricuspid atresia have only a 10% chance of survival beyond the first year of life. The final goal is to perform a univentricular type of repair, with a modified Fontan procedure or total cavopulmonary connection (see chapter "Single ventricle"). The timing and the type of the initial palliation depends upon the amount of antegrade pulmonary blood flow.

In the presence of ductus-dependent pulmonary blood flow, after medical treatment with administration of prostaglandins for stabilization of the clinical condition, a systemic-to-pulmonary artery shunt (modified Blalock-Taussig shunt: see chapter: "Tetralogy of Fallot") is required in the neonatal period.

In the presence of reduced pulmonary blood flow, the majority of infants require a systemic-to-pulmonary artery shunt, and the timing of intervention is correlated with the severity and the progression of the obstruction to the pulmonary blood flow. Nevertheless, about 20% of infants with more balanced pulmonary circulation do not require a systemic-to-pulmonary artery shunt, but are candidate to a cavopulmonary connection (= bidirectional Glenn; see chapter "Single ventricle") as first stage of their surgical treatment.

In the presence of unrestricted antegrade pulmonary blood flow, pulmonary artery banding (see chapter "Ventricular septal defect") is required to reduce the distal pulmo-



nary artery pressure and flow, in order to protect the pulmonary vascular bed and to preserve the function of the left ventricle for a future total cavopulmonary connection (see chapter "Single ventricle").

In the presence of a concordant a ventriculoarterial connection, a potential surgical option utilized in the past, and recently taken again into consideration, thanks to the extended application of the one-and-half ventricular type of repair (see chapter "Ebstein's anomaly"), is the closure of atrial and ventricular septal defects, with end-to-side anastomosis of the superior vena cava to the right pulmonary artery (=bidirectional Glenn) and the connection of the hypoplastic subpulmonary chamber to the pulmonary artery. This surgical approach, utilizing the subpulmonary chamber to pump the return from the inferior vena cava into the pulmonary circulation, has the advantages of the one-and-half ventricular type of repair: pulsatile flow in the pulmonary arteries, low pressure in the right atrium, coronary sinus and splanchnic venous system. The disadvantage of leaving the right atrium within this type of cavopulmonary connection must be balanced against the advantage of incorporating a pumping chamber, even if relatively small, into the right-sided circulation.

In neonates with tricuspid atresia with a discordant ventriculoarterial connection, restrictive ventricular septal defect (therefore with systemic obstruction, with or without associated obstruction at the level of the aortic arch and/or isthmus) and pulmonary hypertension, pulmonary artery banding is contraindicated because it will accelerate the development of subaortic obstruction and the ventricular hypertrophy (contraindica-

Fig. 2.2.4. Tricuspid atresia: angiocardiography. Left ventricular injection showing a ventricular septal defect opacification of a very small subpulmonary right ventricular chamber with ventriculoarterial concordance: **a** anteroposterior view, **b** left anterior oblique view, **c** lateral view (*Ao* aorta, *LPA* left pulmonary artery, *LV* left ventricle, *MPA* main pulmonary artery, *RPA* right pulmonary artery, *RV* right ventricle)

tion for a successful future total cavopulmonary connection). In these cases the surgical treatment consists in a Norwood first stage procedure (see chapter "Hypoplastic left heart syndrome"), converting the patient into one with unobstructed systemic blood flow and pulmonary atresia (treated with either a modified Blalock-Taussig shunt or the Sano modification; see chapter "Hypoplastic left heart syndrome"). Another potential option (less frequently used because of the above reasons) is the surgical enlargement of the ventricular septal defect with or without subaortic resection, to reduce the obstruction to the systemic blood flow, associated with pulmonary artery banding.

Surgical treatment

- *Modified Blalock-Taussig shunt*: see chapter "Tetralogy of Fallot".
- *Pulmonary artery banding*: see chapter "Single ventricle".
- *Bidirectional Glenn* (Figs. 2.2.5 and 2.2.6) *or Hemi-Fontan*: see chapter "Single ventricle".

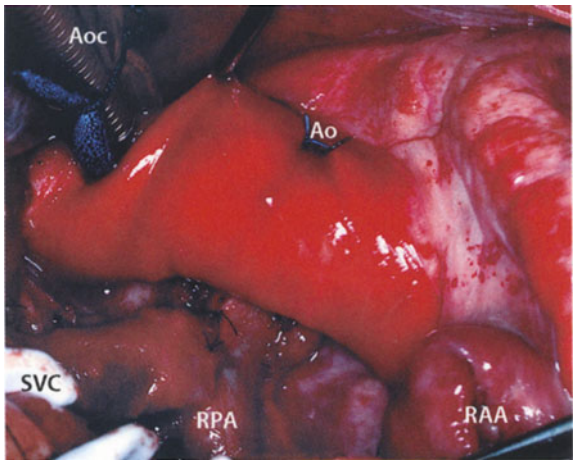


Fig. 2.2.5. Tricuspid atresia: surgery. Bidirectional Glenn. Intraoperative photograph of a bidirectional Glenn, with end-to-side anastomosis of the superior vena cava to the right pulmonary artery, exposed by retraction of the right auricular appendage, after removal of all cardiopulmonary bypass cannulas, with the exception of the aortic cannula (*Ao* aorta, *Aoc* aortic cannula, *RAA* right auricular appendage, *RPA* right pulmonary artery, *SVC* superior vena cava)

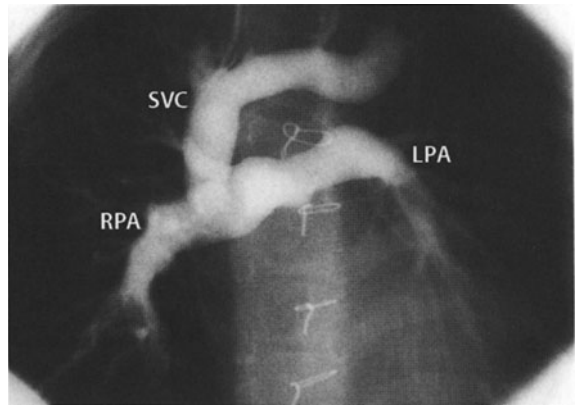


Fig. 2.2.6. Tricuspid atresia: surgery. Post-operative angiography after end-to-side anastomosis of the superior vena cava to the right pulmonary artery (=bidirectional Glenn) in the anteroposterior view with contrast injection in the superior vena cava, showing opacification of both pulmonary arteries (*LPA* left pulmonary artery, *RPA* right pulmonary artery, *SVC* superior vena cava)

- *Modified Fontan procedure or total cavopulmonary connection*: see chapter "Single ventricle".
- *One-and-a-half ventricular repair*: this surgical approach, performed on cardiopulmonary bypass, consists in the confection of a bidirectional Glenn: end-to-side anastomosis between the proximal stump of the superior vena cava transacted at the level of the cavoatrial junction (preserving intact the sinus node, and its artery) and the upper aspect of the right pulmonary artery, opened with either a longitudinal or transversal incision (according to the surgeon's preference). The atrial and ventricular septal defects, respectively, approached through a right atriotomy and a longitudinal incision of the subpulmonary chamber, are closed with separated patches (pericardium, PTFE, Dacron or Teflon), and then the right atrium is connected with the subpulmonary chamber with interposition of a valved conduit. In the presence of associated pulmonary valve stenosis, a pulmonary valvotomy is required. In the presence of a hypoplastic pulmonary valve annulus, the incision of the subpulmonary chamber is prolonged becoming a transannular opening, and the distal end of the conduit

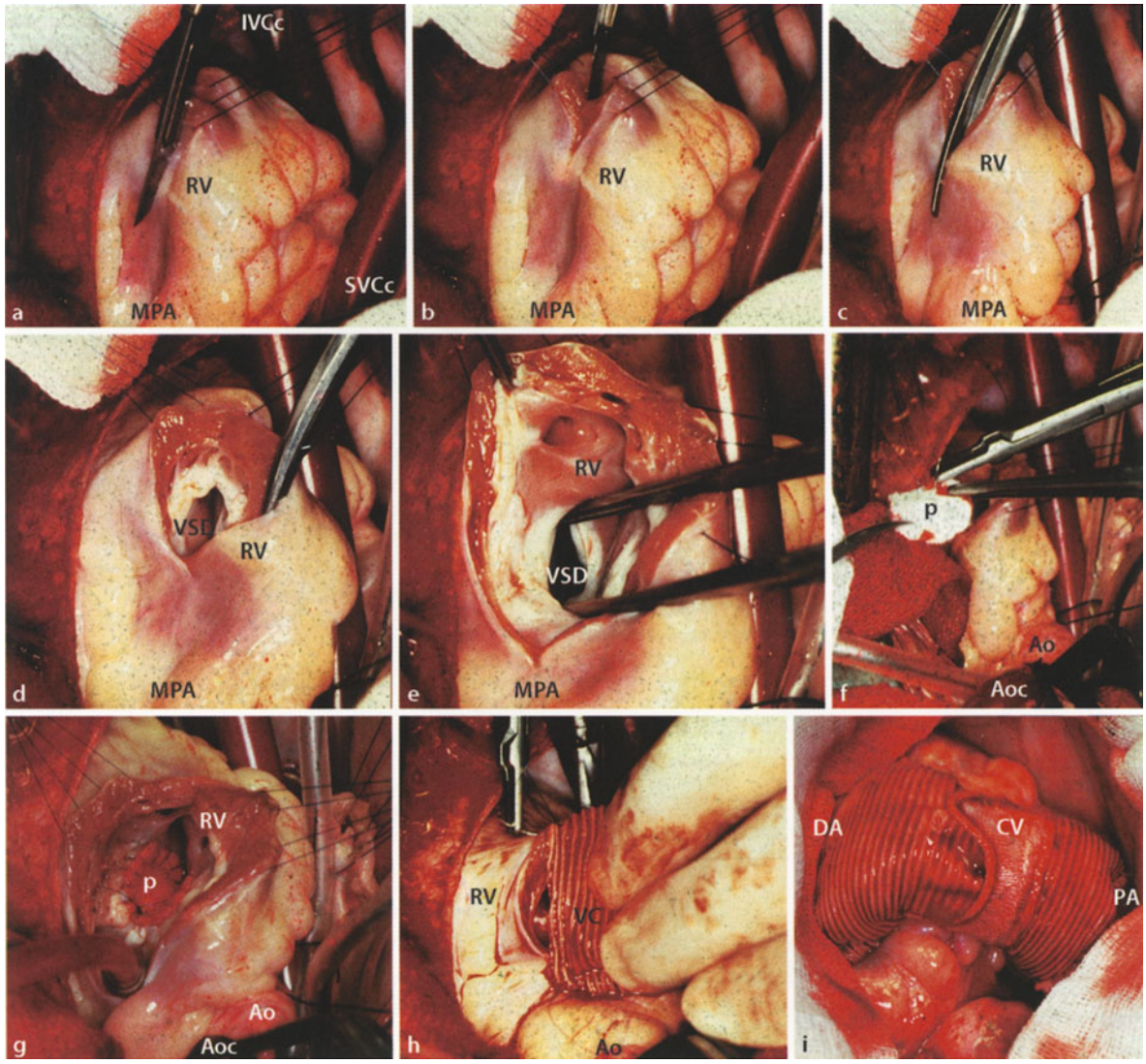


Fig. 2.2.7. Tricuspid atresia: surgery. Intraoperative photograph of one-and-a-half ventricular repair. **a** Preparation with stay sutures for longitudinal incision of the small subpulmonary right ventricular chamber in the direction of the pulmonary artery (*IVCc* inferior vena cava cannula, *MPA* main pulmonary artery, *RV* right ventricle, *SVCc* superior vena cava cannula), **b** initial longitudinal incision of the small subpulmonary right ventricular chamber in the direction of the pulmonary artery with the knife, **c** extension with scissors of the longitudinal incision of the small subpulmonary right ventricular chamber in direction of the pulmonary artery, **d** initial exposure of the ventricular septal defect, with a fibrous edge, through the longitudinal incision of the small subpulmonary right ventricular chamber (*VSD* ventricular septal defect), **e** complete exposure of the ventricular septal defect through the longitudinal incision of the small subpulmonary right ventricular chamber, **f** initial patch closure of the ventricular septal defect (*Ao* aorta, *Aoc* aortic cannula, *p* patch), **g** completed patch closure of the ventricular septal defect, **h** distal anastomosis of a Dacron conduit with a biological valve on the edge of the right ventriculotomy (*VC* valved conduit), **i** completed implantation of the Dacron conduit with a biological valve from the right atrium to the subpulmonary right ventricular chamber (*CV* conduit valve, *DA* distal anastomosis, *PA* proximal anastomosis)

is anastomosed to both the subpulmonary chamber and the wall of the pulmonary artery (Fig. 2.2.7).

- *Subaortic resection*: the technique, on cardiopulmonary bypass, consists of a longitudinal incision of the subaortic right ventricular outlet chamber in the direction of the ascending aorta, of course avoiding major coronary artery branches. After careful identification of the ventricular septal defect (=bulboventricular foramen), a full-thickness of interventricular septum is resected from the anterosuperior aspect of the defect, on the opposite side of the conduction tissue, carefully avoiding lesions to the adjacent aortic valve. Subaortic resection is completed with excision of obstructing muscle bundles, and the outlet chamber is further enlarged by patch (autologous or heterologous pericardium, PTFE) closure of the incision.

Potential complications

- *Modified Blalock-Taussig shunt*: see chapter "Tetralogy of Fallot".
- *Pulmonary artery banding*: see chapter "Single ventricle".
- *Bidirectional Glenn or Hemi-Fontan*: see chapter "Single ventricle".
- *Modified Fontan procedure or total cavopulmonary connection*: see chapter "Single ventricle".
- *Subaortic resection*: complete atrioventricular block should occur very rarely, while the risk for either residual or recurrent subaortic obstruction is higher.

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Chapter 2.3 Single ventricle

Incidence

Single ventricle is reported with an incidence varying between 0.87 and 1.27 in 10,000 newborns. No sex prevalence is reported.

Morphology (Fig. 2.3.1)

Single ventricle (or univentricular heart) is considered a congenital cardiac malformation in which both atria connect to only one ventricular chamber by either two separate atrioventricular valves (double inlet) or a common atrioventricular valve (common inlet). The ventricle to which both atrioventricular valves or a common atrioventricular valve connects is generally well formed, whereas the ventricle not receiving the largest amount of the venous return to the heart is often a rudimentary chamber.

By definition, the term double inlet single ventricle is used only if more than 50% of the overriding valve lies over the main ventricular chamber. When both atrioventricular valves are present, they often cannot be designated as either mitral or tricuspid, and are commonly straddling or stenotic. When there is a common atrioventricular valve, it presents frequently with regurgitation.

Most of the hearts described as "single ventricle" or "univentricular heart" in reality possess two ventricular chambers, with one main (dominant) chamber and a second (incomplete) rudimentary chamber which lacks one or more of its components (generally the inlet, but occasionally also the outlet).

There are three basic patterns of ventricular morphology:

- main ventricular chamber of left ventricular morphology, with rudimentary right ventricle (= single ventricle of left ventricular type),
- main ventricular chamber of right ventricular morphology, with or without a rudimentary left ventricle (= single ventricle of right ventricular type),
- single ventricular chamber of indeterminate morphology.

The combination of the type of inlet (double inlet or common inlet) with the type of ventricular morphology (left, right or indeterminate) provide a variety of univentricular atrioventricular connections. This variety is further more complicated when other morphological variables are taken in consideration, like the cardiac position (levocardia, mesocardia or dextrocardia), the atrial situs (solitus, inversus, right or left isomerism), the ventriculoarterial connections, the spatial relationship between the main chamber and the rudimentary chamber, and the associated cardiac anomalies.

The most frequent arrangement is the main chamber of left ventricular morphology (Figs. 2.3.2 and 2.3.3) with a rudimentary chamber of right ventricular morphology (= single ventricle of left ventricular type).

The rudimentary chamber is separated from the main chamber by a septum that does not extend to the crux of the heart, and is connected to the main chamber via a ventricular septal defect, variously described

A. F. Corno, *Congenital Heart Defects*

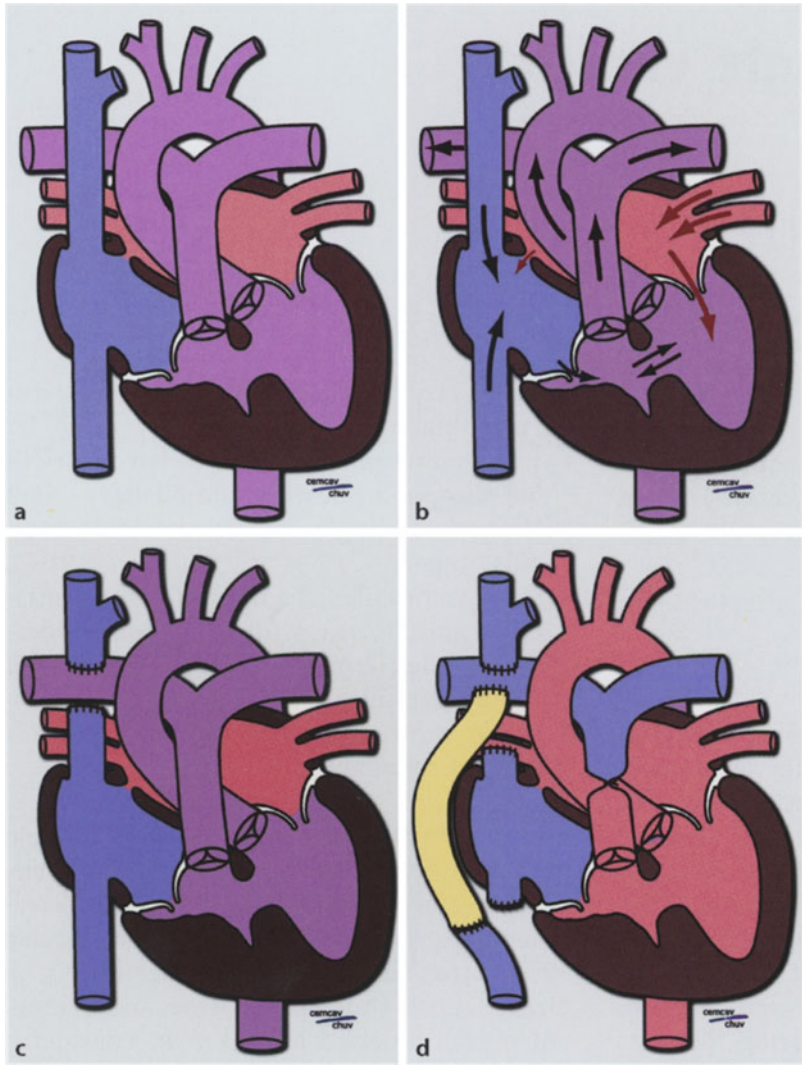

Fig. 2.3.1. Single ventricle: **a** morphology, **b** pathophysiology, and surgery: **c** superior vena cava to right pulmonary artery anastomosis, and **d** extracardiac total cavopulmonary connection

with interchangeable terms as outlet foramen, interventricular foramen, or bulboventricular foramen. Since these hearts do not have a membranous septum, the ventricular septal defect is generally of completely muscular type, with the potential reduction in size typical of the muscular ventricular septal defect in biventricular hearts. In order to evaluate the size of the ventricular septal defect, and to define whether its size is restrictive or unrestrictive, generally its dimensions are related to the size of the corresponding aortic root.

The second chamber is of right ventricle morphology, is always anterior, and is located either to the left or the right. The size of the outlet chamber is related to the degree of development and straddling of the tricuspid valve, in addition to the size of the ventricular septal defect. The ventriculoarterial connections are most commonly discordant and more rarely concordant. Unusual forms of ventriculoarterial connections in double inlet ventricle include double outlet or single outlet (=pulmonary atresia).

Outflow obstruction to the pulmonary artery is common, and is the most important determinant of the clinical course. The obstruction may be subvalvular and/or valvular (hypoplastic annulus and/or thickened leaflets), or may be complete (=pulmonary atresia). In the presence of ventriculoarterial

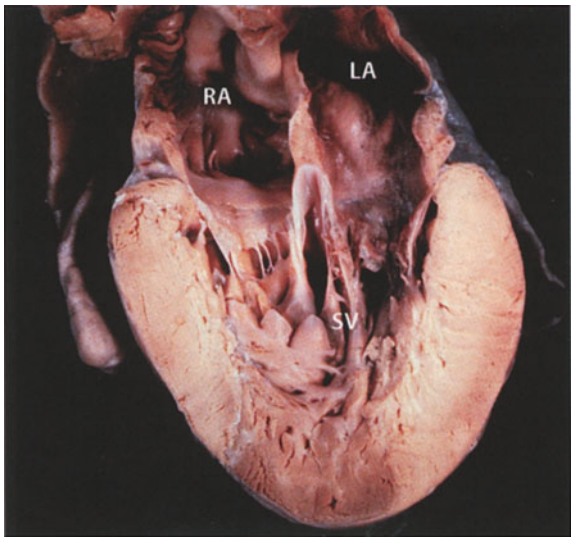

Fig. 2.3.2. Single ventricle: morphology. Specimen of a heart with double inlet single ventricle of left ventricular morphology, with complete absence of an interventricular septum between the two atrioventricular valves (*LA* left atrium, *RA* right atrium, *SV* single ventricle) (reproduced with permission from Marino B, Thiene G (1990) Atlante di anatomia ecocardiografica delle cardiopatie congenite, USES, Firenze)

concordance the obstruction, mostly dynamic, due to infundibular narrowing, is generally localized at the subvalvular level. In hearts with ventriculoarterial discordance the obstruction of the pulmonary blood flow is generally due to the presence of a restrictive ventricular septal defect.

Obstruction to the systemic outflow can occur at the subvalvular level and/or the level of the aortic arch and/or isthmus, or at multiple levels. Usually it occurs at the level of the ventricular septal defect in hearts with discordant ventriculo-arterial connections; in fact, in these patients the presence of aortic coarctation and/or aortic arch hypoplasia is a strong marker for the presence of a restrictive ventricular septal defect.

The conduction tissues, as seen in relationship to the ventricular septal defect, have a directly comparable arrangement to that seen in tricuspid atresia. Unlike tricuspid atresia, however, the AV node is situated anteriorly within the right atrioventricular orifice rather than within the atrial septum. This arrangement is seen irrespective of whether the rudimentary right ventricle is right or left-sided. The position of the rudimentary ventricle affects only the relationship of the atrioventricular bundle to the outflow tract from the dominant left ventricle. From the surgical point of view, the atrioventricular node can be anywhere around the perimeter of the right-sided (=draining the right-sided atrium) atrioventricular valve.

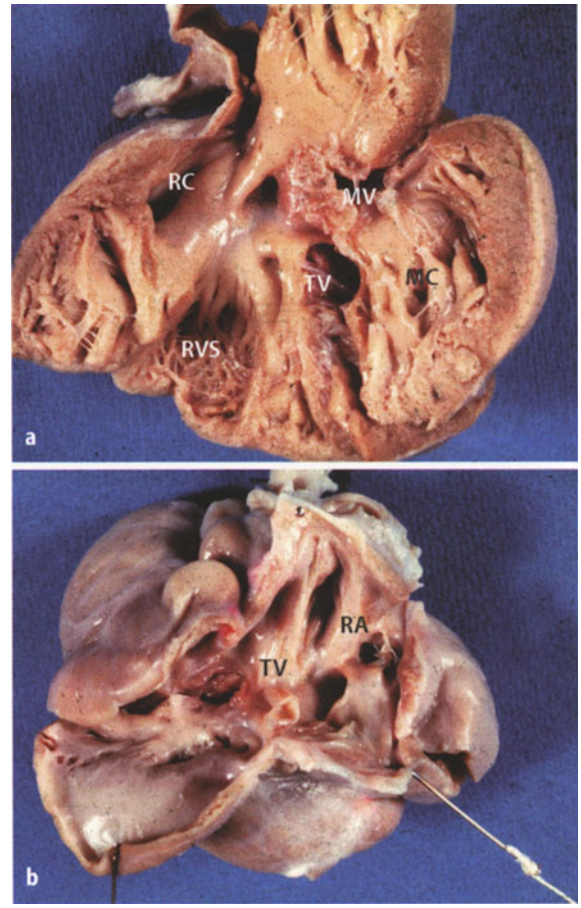

Fig. 2.3.3. Single ventricle: morphology. **a** Specimen of a heart with dextrocardia, single ventricle of left ventricular morphology, with double inlet because of the presence of mitral valve and imperforated tricuspid valve, and a blind sinus of right ventricular morphology (*MC* main chamber, *MV* mitral valve, *RC* rudimentary chamber, *RVS* right ventricular sinus, *TV* tricuspid valve), **b** specimen of the same heart as in **a** showing the right atrium with imperforated tricuspid valve (*RA* right atrium) (reproduced with permission from Corno AF, Casolo F, Fancini P, Guffanti MC, Thiene G (1979) Coesistenza di cuore univentricolare sinistro con seno ventricolare destro. Progr Cardiol Pediat 157–162)

Associated anomalies

Associated cardiac anomalies occur in about one-third of cases with double inlet. Anomalous pulmonary and/or systemic venous connection are relatively frequent, particularly in patients with atrial isomerism (see chapter "Isomerism"); the most frequent are persistent left superior vena cava, interruption of the inferior vena cava, and partial or total anomalous pulmonary venous connection. Malformations of the atrioventricular valves are also quite common, including straddling, leaflet dysplasia, leaflet cleft and tags, and annular hypoplasia. Aortic arch anomalies such as coarctation, interrupted aortic arch and hypoplastic aortic arch are strongly associated with a restrictive ventricular septal defect in cases in which the aorta arises from the rudimentary chamber (=ventriculoarterial discordance).

Pathophysiology

Functional definition of a single ventricle includes different morphological conditions with one of the two ventricles too small to allow for survival with the two circulations in series: desaturated systemic venous return pumped by a ventricle to the pulmonary circulation and oxygenated pulmonary venous return pumped by the other ventricle to the aorta. This definition of "functionally univentricular hearts", widely used in the literature of last few decades, includes malformations like tricuspid atresia and mitral atresia, not considered in this chapter.

Lack of a separation between the pulmonary and systemic circulations causes obvious cyanosis, with the severity depending upon the degree of obstruction to the pulmonary blood flow. There are several important hemodynamic variables in single ventricle, including the degree of mixing at the atrial level (poor, moderate or complete), the degree of outlet obstruction, and the presence of atrioventricular valve malfunction.

In single ventricle, mixing of systemic and pulmonary venous return occurs in the main ventricular chamber; this may result in nearly equal oxygen saturation in the aorta and the pulmonary artery (*complete mixing*). In some cases, streaming of blood within the ventricle results in a substantial difference in oxygen saturation between the aorta and the pulmonary artery. The *streaming* may be *favorable*, with aortic saturation being greater than pulmonary arterial saturation, or *unfavorable*, with pulmonary saturation being greater than aortic (=hemodynamics of transposition of the great arteries).

The degree of saturation is also influenced by the presence and severity of pulmonary stenosis, the single most important determinant of the clinical course. In the absence of obstruction to the pulmonary blood flow, the presence of systemic obstruction is also important, although less common, and almost always due to subvalvular stenosis at the level of the ventricular septal defect or bulboventricular foramen, more or less associated with hypoplastic aortic arch, aortic coarctation or aortic arch interruption.

Severe atrioventricular valve stenosis or regurgitation is hemodynamically important if the atrial septum is intact or the atrial septal defect is restrictive, in which case the venous return may be restricted or obstructed.

An important point to consider regarding the pathophysiology of hearts with a single ventricle is the transformation in form and function they may undergo over time. These changes are related to modifications at the following levels:

- effective size of the ventricular septal defect
- progression in the obstruction to either the systemic or the pulmonary blood flow
- progression of the atrioventricular valve(s) regurgitation
- progressing ventricular hypertrophy, with the subsequent reduced ventricular compliance and impaired diastolic function
- impaired systolic ventricular function

- progressive atrioventricular conduction disturbances, including complete atrioventricular block
- progressive left atrial hypertension, consequent to restrictive interatrial communication and/or obstructed atrioventricular junction
- progress towards pulmonary vascular obstructive disease

Diagnosis

Clinical pattern: there are different patterns of presentation, occurring in the neonatal period or during early infancy, depending upon the amount of pulmonary blood flow;

- neonates with ductus-dependent pulmonary blood flow because of the presence of pulmonary atresia, typically present shortly after birth with severe cyanosis, while neonates with reduced pulmonary blood flow due to pulmonary stenosis generally present with cyanosis within the first week of life;
- children with increased pulmonary blood flow usually present within the first few months of life;
- patients with balanced pulmonary blood flow may present much later in life with only mild cyanosis;
- cyanosis is more evident in cases with obstructed pulmonary blood flow, while congestive cardiac failure is more common in those with unrestricted pulmonary blood flow;
- severe atrioventricular valve regurgitation results in elevated atrial pressure and the early appearance of congestive heart failure;
- systemic obstruction is accompanied by the signs of poor peripheral perfusion, with weak peripheral pulses, rapid breathing, lethargy, poor feeding, oliguria, metabolic acidosis;
- the second heart sound is generally single and accentuated; outlet obstruction is accompanied by systolic ejection murmur.

Electrocardiogram: R/S or r/S pattern from V4R to V6 is suggestive of a single ventricle; septal Q wave in the right precordial leads in cases with a single ventricle of the left ventricular type; qR pattern in the right precordial leads and r/S in the left precordial leads in cases with a single ventricle of the right ventricular type.

Chest X-ray: variable accordingly with the hemodynamic pattern, particularly with the presence and degree of obstruction to the pulmonary blood flow; cardiomegaly is associated with markedly increased pulmonary blood flow or with substantial atrioventricular valve regurgitation or with important ventricular dysfunction; severe pulmonary edema is associated with an obstructed total anomalous pulmonary venous connection or with restrictive interatrial communication in the presence of stenosis of the left atrio-ventricular valve.

Echocardiogram: most of the main features of a single ventricle can be defined by echocardiography with a combination of apical 4-chamber and parasternal long-axis views and precordial and subcostal 4-chamber and short-axis views (Figs. 2.3.4–2.3.6), in partic-

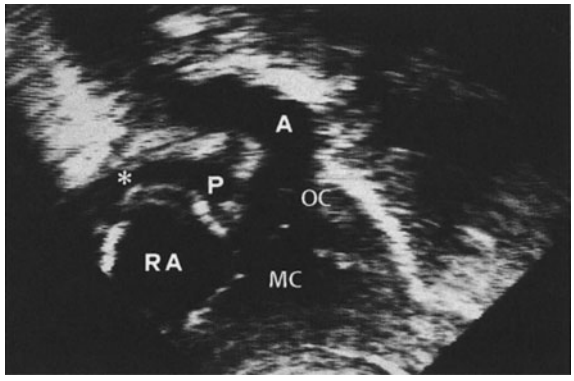

Fig. 2.3.4. Single ventricle: echocardiography. Oblique right subxiphoid view showing the connection between the right atrium and the single ventricular chamber, with obstruction to the pulmonary outflow and small right pulmonary artery (asterisk) (*A* aorta, *MC* main ventricular chamber, *OC* outlet ventricular chamber, *P* pulmonary artery, *RA* right atrium) (reproduced with permission from Marino B, Thiene G (1990) Atlante di anatomia ecocardiografica delle cardiopatie congenite, USES, Firenze)

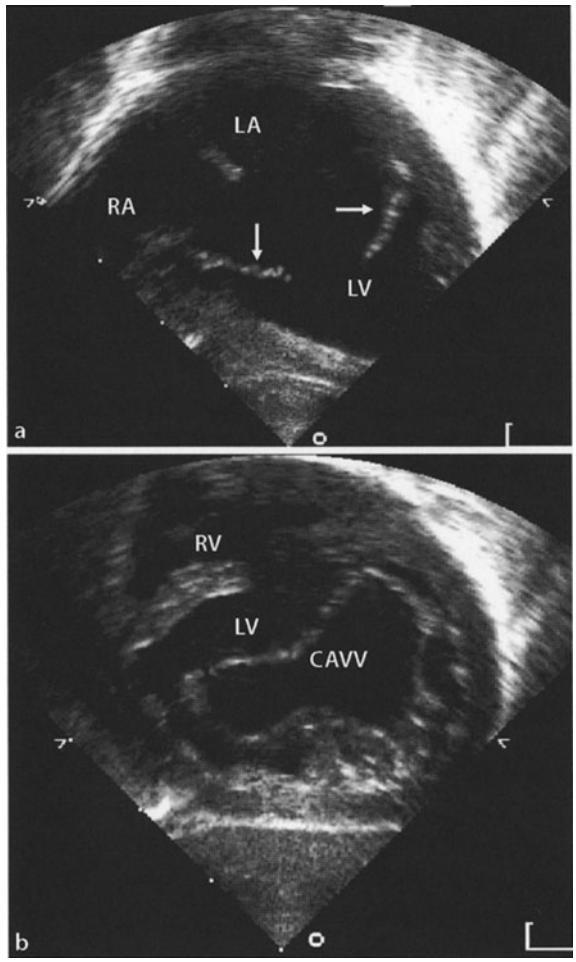

Fig. 2.3.5. Single ventricle: echocardiography. **a** 4-chamber view showing a single ventricle of the left ventricular type with a common atrioventricular valve (white arrows) (*LA* left atrium, *LV* left ventricle, *RA* right atrium), **b** short-axis view in the same patient showing the single ventricle of the left ventricular type with the common atrioventricular valve (*CAVV* common atrioventricular valve, *LV* left ventricle, *RV* right ventricle) (photographs courtesy of Dr. Michael Rigby)

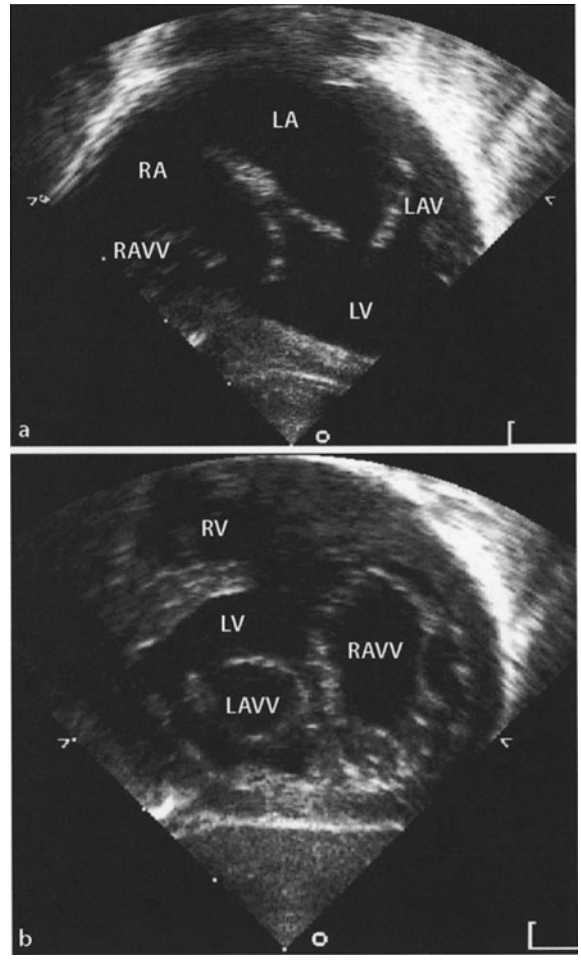

Fig. 2.3.6. Single ventricle: echocardiography. **a** 4-chamber view showing a double inlet single ventricle of left ventricular type (*LA* left atrium, *LAVV* left atrioventricular valve, *LV* left ventricle, *RA* right atrium, *RAVV* right atrioventricular valve), **b** short-axis view in the same patient showing a double inlet single ventricle of the left ventricular type (photographs courtesy of Dr. Michael Rigby)

ular the presence and degree of subpulmonary obstruction, the presence and degree of systemic obstruction (subaortic, at the level of aortic arch or aortic isthmus), and the presence and degree of atrio-ventricular valve regurgitation.

Cardiac catheterization: in most case it is utilized to integrate the information provided by echocardiography, particularly regarding the hemodynamics; important left atrioventricular valvular stenosis should be determined, as its presence necessitates balloon atrial septectomy (=Rashkind procedure); pulmonary arterial pressures and resistance must be evaluated, since the choice of the operative therapy is largely dictated by the presence and degree of pulmonary stenosis or pulmonary hypertension; the presence and degree of subaortic obstruction must be evaluated, sometimes with pharmacological intervention (isoprenaline administration) to "unmask" dynamic subaortic obstruction.

Indications for surgical treatment

The estimated overall survival without treatment of patients born with a single ventricle is about 57% at 1 year and 45% at 5 years, although some subjects with balanced circulations may have more favorable prognosis.

Medical management is largely restricted to the management of congestive cardiac failure in cases with unrestricted pulmonary blood flow, or to the use of prostaglandins in neonates with ductus-dependent pulmonary blood flow because of critical pulmonary stenosis or pulmonary atresia, or with ductus-dependent systemic distal perfusion because of severe obstruction or interruption of the aortic arch.

Occasionally, percutaneous balloon or blade atrial septostomy is required to relieve pulmonary venous obstruction in cases with left-sided atrioventricular valve stenosis and an intact atrial septum or restrictive atrial septal defect.

Ideally, surgical treatment consists of complete separation of the systemic and pulmonary circulation.

Fontan type circulation

The surgical treatment is generally directed by the ultimate goal of achieving a Fontan type of circulation, with complete separation of the systemic from the pulmonary circulation, by deviating the desaturated systemic venous return from the superior and inferior vena cava directly to the pulmonary arteries (without a pumping chamber), and by using the single ventricle to pump the oxygenated pulmonary venous return to the aorta. The Fontan type of circulation, being highly dependent on the presence of key physiological conditions, requires a plan of staged-reconstruction in order to obtain and maintain several features necessary for the adequate functioning of the Fontan circulation:

- the single ventricle, the only pumping chamber available, needs to have adequate systolic (=contractility) (ejection fraction not lower than 45%) and diastolic functions (=compliance) (end-diastolic ventricular pressure not higher than 15 mmHg in the absence of severe atrioventricular valve regurgitation); therefore either volume overload as well as pressure overload have to be avoided, particularly in the long term;
- the atrioventricular valve(s) must be preserved or repaired if needed, since either atrioventricular valve regurgitation or stenosis can impair the ventricular function on the one hand and present an obstacle to the pulmonary venous return on the other hand,
- any systemic obstruction (either subaortic or at the level of the aortic arch and/or isthmus) must be relieved,
- the pulmonary arteries must grow with normal size (McGoon index higher than 0.75) and morphology, without stenosis and/or distortion, and with low pulmonary artery pressure (mean pressure lower than 15–20 mmHg) and pulmonary vascular resistance (less than 4 units/m^2 of BSA), in order to allow unimpeded blood flow from the superior and vena cava through the lungs,
- the normal sinus rhythm must be preserved, because absence of sinus rhythm reduces the ventricular performance.

Staged-reconstruction consists of three stages, the first being a palliative procedure in which the systemic and pulmonary circulations are usually placed in parallel, the second stage consisting of a superior cavopulmonary anastomosis (=bidirectional Glenn), and the final stage being conversion to a total cavopulmonary connection (Fontan physiology). The QP:QS (=pulmonary-to-systemic blood flow ratio) for the three stages are typically 2–3:1, 0.5:1, and 1:1.

Infants with restricted pulmonary blood flow and no subaortic stenosis or left ventricular outflow tract obstruction are treated with a systemic-to-pulmonary artery shunt. Pulmonary artery reconstruction may or may not be required depending on the presence of discontinuous pulmonary arteries, pulmonary artery stenosis or distortion, or significant aortopulmonary collaterals.

Infants with a univentricular atrioventricular connection and left ventricular outflow tract obstruction are considered for a modified Damus-Kaye-Stansel procedure (see chapter "Transposition of the great arteries") in which the two great arteries are connected proximally, and the pulmonary blood flow is provided by a systemic-pulmonary shunt (first stage), followed by a superior cavopulmonary anastomosis (=bidirectional Glenn, second stage), and then by a total cavopulmonary connection (=modified Fontan procedure, third stage).

If the left ventricular outflow tract obstruction extends into the aortic arch (hypoplastic aortic arch) or aortic isthmus and descending thoracic aorta (aortic coarctation), a Norwood procedure (see chapter "Hypoplastic left heart") is required in order to relieve the distal systemic obstruction.

Finally, the arterial switch operation has also been utilized, transforming the subaortic stenosis to subpulmonary stenosis and thereby theoretically protecting the pulmonary vascular bed. Many of the cases undergoing the arterial switch operation for a univentricular atrioventricular connection ultimately require a systemic-to-pulmonary artery shunt because of the inadequate pulmonary blood flow, reducing the possibilities of an ultimate Fontan-type operation.

Patients with unrestricted pulmonary blood flow and without subaortic stenosis or left ventricular outflow tract obstruction may initially be palliated with pulmonary artery banding. Drawbacks to this procedure are the potential for pulmonary artery distortion, development of pulmonary valvular regurgitation or damage to the pulmonary valve, progressive ventricular hypertrophy, decreased ventricular compliance, and development of subvalvular aortic stenosis. The latter may result from both ventricular hypertrophy and geometric changes to the left ventricle. Pulmonary artery banding may therefore increase the risk of a subsequent Fontan-type procedure.

In patients coming to observation too late, with severe pulmonary hypertension and a huge and dilated pulmonary artery, an open lung biopsy (Fig. 2.3.7) may be necessary to quantitate the pulmonary vascular lesions in order to facilitate the following decision-making process with regard to the best palliative treatment (progressive pulmonary artery banding, palliative atrial or arterial switch to improve the oxygenation, or other).

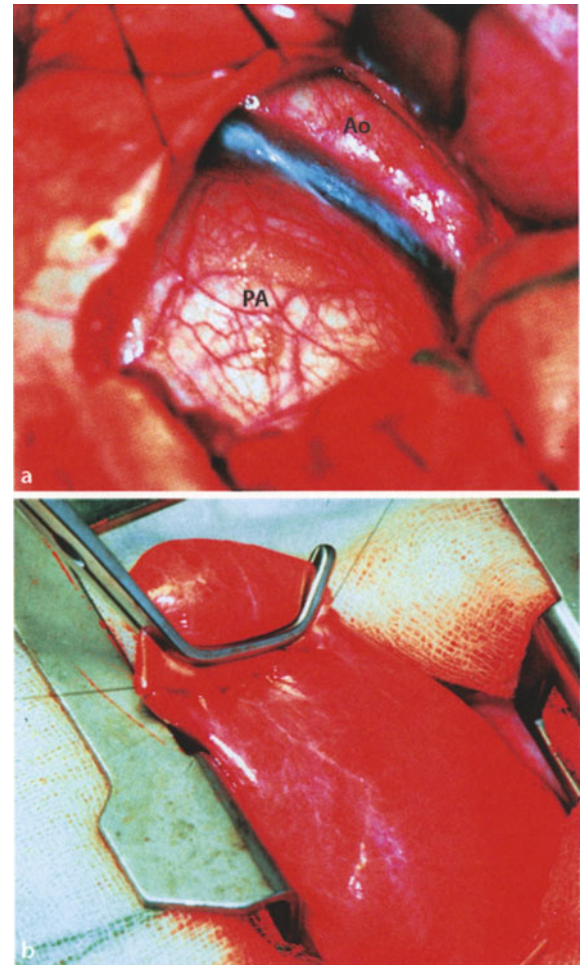

Fig. 2.3.7. Single ventricle. **a** Intraoperative photograph of a patient with late referral, with severe pulmonary hypertension, and a very dilated pulmonary artery, showing the ratio between the diameter of the two great arteries, (*Ao* aorta, *PA* pulmonary artery), **b** intraoperative photograph of the same patient showing the open lung biopsy of the lingual, controlled with a vascular clamp under inflation with positive pressure ventilation

Direct surgical enlargement of the ventricular septal defect with enlargement or resection of the infundibular septum may be required depending on the degree of restric-

tion of the subaortic stenosis. Atrial septectomy may be required in cases with stenosis of the left atrioventricular valve or those with an intact or nearly intact atrial septum.

The second stage operation, the bidirectional Glenn procedure, consists of the division of the superior vena cava corresponding with the cavoatrial junction (preserving the sinus node and its artery) and its end-to-side anastomosis to the upper aspect of the right pulmonary artery.

A bilateral bidirectional Glenn procedure (= end-to-side anastomosis, respectively, of the right superior vena cava to the right pulmonary artery and of the left superior vena cava to the left pulmonary artery) is required in patients with persistent left superior vena cava. The bidirectional Glenn procedure can generally be performed at 4–6 months of life, when the pulmonary vascular resistance is usually sufficiently low to accommodate approximately one third of the systemic venous return in addition to the antegrade pulmonary blood flow. Insufficient pulmonary blood flow may be increased by the addition of a small systemic-to-pulmonary artery shunt.

The advantages of the bidirectional Glenn procedure are the following:

- reduced ventricular volume overload, by reducing the systemic venous return of 35–40% (the return from the superior vena cava),
- reduced blood flow through the atrio-ventricular valve(s),
- increased total pulmonary blood flow and effective pulmonary blood flow, by deviating the most desaturated blood (the superior vena cava return) directly through the pulmonary arteries, thereby reducing the cyanosis,
- avoid the adaptation of the single ventricular chamber to the suddenly reduced ventricular filling required for the single stage modified Fontan procedure, with the subsequent reduced ventricular compliance and poor functioning of the Fontan circulation,
- preparation for later completion of the total cavopulmonary connection, without need for any period of myocardial ischemia,
- being a direct anastomosis between two native vessels, it has the potential for growing with the child.

A surgical alternative to the bidirectional Glenn procedure is the Hemi-Fontan operation, consisting in the connection of the superior vena cava and the superior portion of the right atrium to both pulmonary arteries, augmentation of the central pulmonary arteries, occlusion of the inflow of the superior vena cava to the right atrium and elimination of the other sources of pulmonary blood flow.

Hemi-Fontan operation:

- *advantages:* the hemodynamics obtained is exactly the same as after a bidirectional Glenn procedure, with the only difference being that in this case all the other sources of pulmonary blood flow are eliminated; this could be seen as an advantage with regard to the ventricular volume overload, but a potential disadvantage in patients with pulmonary arteries of relatively small size because of suboptimal pulmonary artery growth, since only 40% of the systemic venous return perfuses the pulmonary circulation until the completion of the Fontan operation; this potential disadvantage should be compensated for by the direct enlargement of the central pulmonary arteries;
- *disadvantages:* the procedure requires cardiopulmonary bypass with aortic cross clamp (therefore a period of myocardial ischemia), biological or prosthetic materials for central pulmonary arteries augmentation, suturing lines in correspondence or in the proximity of the sinus node or its artery (with the relative risk of early or late supraventricular arrhythmias), and it offers the possibility for later conversion to a total cavopulmonary connection with a lateral tunnel technique (sometimes even without cardiopulmonary bypass), but it complicates the possibility of an extracardiac inferior vena cava to pulmonary artery connection.

Conversion to a Fontan operation (total cavopulmonary connection) completes the

single-ventricle pathway in cases with unrestricted pulmonary blood flow.

Total cavopulmonary connection nowadays is achieved with one of the two following techniques: a lateral tunnel or an extracardiac inferior vena cava to the pulmonary artery connection. Both the lateral tunnel and the extracardiac connection present the same advantages as the abandoned techniques of atriopulmonary connection:

- the hemodynamics offers substantial improvement in terms of energy conservation, thanks to reduced flow turbulence,
- both techniques are easy to reproduce and suitable for all types of atrioventricular valve arrangements,
- the coronary sinus remains in the lower pressure left atrial chamber.

Advantages of the extracardiac total cavopulmonary connection compared with the interatrial lateral tunnel technique:

- no myocardial ischemic period is needed, because it can be performed on the beating heart, therefore without aortic cross clamping, and even without cardiopulmonary bypass,
- hemodynamics benefits due to the maximized laminar flow principle,
- no need for intraatrial incisions and sutures, with a subsequent reduction of supraventricular arrhythmias
- applicability to situations with anomalous pulmonary and/or systemic venous connection, with subsequent reduction of obstruction to the pulmonary and/or systemic venous return,
- the fenestration can be easily performed off-bypass.

A relative disadvantage of the extracardiac connection is that, in order to implant an extracardiac conduit of adequate size (18–20 mm diameter), the ideal age for the operation is slightly higher than that for the lateral tunnel technique.

Both of the above techniques can be performed with the addition of a *fenestration*, consisting in an incomplete atrial partitioning, or connection between the right (systemic) and the left (pulmonary) venous channels allowing for a decompression of the systemic venous system (reducing the venous pressure that is too high) and an increase of the ventricular filling (increasing the systemic cardiac output), with the price of systemic oxygen desaturation (because of the iatrogenic right-to-left shunt). The *fenestration* is added to the total cavopulmonary connection either on indication (in higher risk candidates) or routinely, accordingly with the surgical preference.

A further modification of the total cavopulmonary connection is possible in the high-risk Fontan candidate, with the connection of the superior vena cava to the left pulmonary artery, and either an interatrial lateral tunnel with adjustable atrial septal defect or an extracardiac connection between the inferior vena cava and the right pulmonary artery with a fenestration; in this way the lesser systemic venous return (35–40%) from the superior vena cava is deviated into the smaller lung (the left), while the venous return from the inferior vena cava is deviated into the larger lung (the right), but with a potential decompression (adjustable atrial septal defect or fenestration) allowing the maintenance of adequate cardiac output and avoiding venous hypertension in the hepatic and renal systems.

Damus-Kaye-Stansel procedure

In the presence of subaortic obstruction, the surgical possibilities are Damus-Kaye-Stansel procedure (end-to-side or double-barrel anastomosis), or subaortic resection or bulboventricular foramen enlargement (see chapter "Tricuspid atresia"), or arterial switch (see chapter "Transposition of the great arteries") in the neonatal period.

The Damus-Kaye-Stansel procedure (see chapter "Transposition of the great arteries"):

- *advantages*: better relief of the systemic outflow obstruction, lower incidence of complete atrioventricular block, lower incidence of need for reoperation,
- *disadvantages*: increased progression of semilunar valve regurgitation, technically

more demanding, not suitable during ventricular septation.

Ventricular septation

Ventricular septation can be also a viable option in 20% of patients, depending on the ventricular morphology, on the one hand, and on local practice and philosophy on the other hand. Ventricular septation may be an option in patients with an enlarged dominant ventricle, with two atrioventricular valves with little or no overriding or straddling. The ventriculoarterial connection must be amenable to repair with the appropriate ventriculoarterial connection, and there should be little or no pulmonary or systemic outflow obstruction.

This procedure can be performed as a single stage or as a two-stage treatment; this last approach has the purpose of obtaining an early partial separation of the systemic and pulmonary circulations in the first few months of life, leaving a small residual intracardiac shunt to close later. Two-stage ventricular septation remains a technique very rarely utilized.

Heart transplantation

Heart transplantation is considered for patients who are poor candidates for a Fontan-type procedure because of poor ventricular function but where pulmonary vascular resistance is not elevated, or for patients with a failing Fontan procedure not suitable for Fontan take-down or other more conventional surgical treatment.

As a final comment on the indication for surgical treatment, it is highly unlikely that a single management strategy will be optimal for this very heterogeneous group of patients.

Surgical treatment

Pulmonary artery banding

In these patients the achievement of an adequate pulmonary artery banding is particularly difficult, in consideration of the several interrelated variables in continued evolution, particularly the need to obtain and maintain a low pulmonary artery pressure, an adequate balance between the systemic and pulmonary circulation, at the same time avoiding an excessive ventricular pressure overload. A solution to these requirements is the pulmonary artery banding with a telemetrically adjustable device (FloWatch-PAB) (Figs. 2.3.8 and 2.3.9).

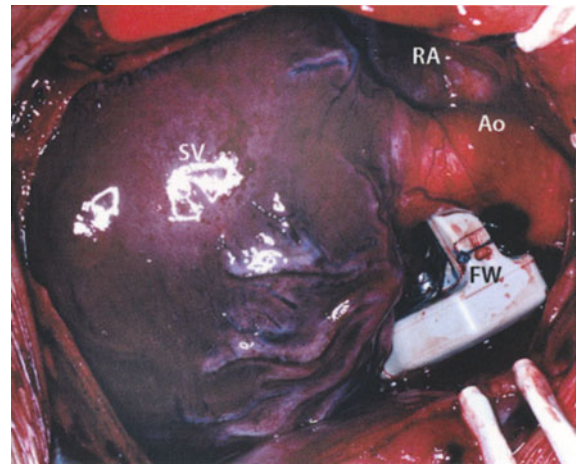

Fig. 2.3.8. Single ventricle: surgery. Palliation: pulmonary artery banding. Pulmonary artery banding with a telemetric adjustable device (FloWatch-R-PAB^{TM}): the device has already been positioned and clipped around the pulmonary artery, posterior to and on the left side of the anterior aorta (*Ao* aorta, *FW* FloWatch-PAB^{TM}, *RA* right atrium, *SV* single ventricle)

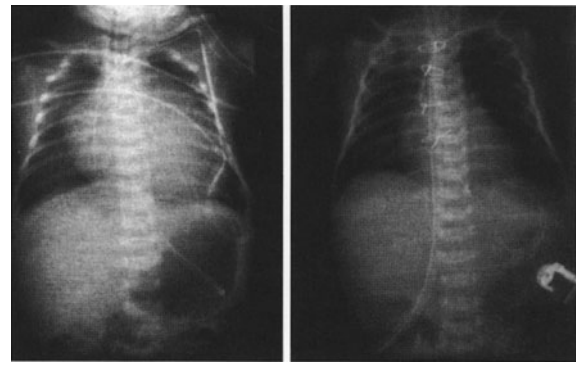

Fig. 2.3.9. Single ventricle: surgery. Palliation: pulmonary artery banding. Chest X-ray in the anteroposterior view showing the substantial reduction of the cardiomegaly immediately obtained with pulmonary artery banding in a neonate with a single ventricle and increased pulmonary artery flow and pressure. *Left:* Preoperative chest X-ray, *right:* postoperative chest X-ray (immediately after surgery)

Shunt

A modified Blalock-Taussig shunt (see chapter "Tetralogy of Fallot") is always the choice for a systemic-to-pulmonary artery shunt.

Bidirectional Glenn (Fig. 2.3.10)

The bidirectional Glenn procedure consists of the ligature and division of the azygos vein to fully mobilize the superior vena cava and to avoid later decompression of the superior vena cava into the inferior vena cava. Then the superior vena cava, controlled with a vascular clamp, is transected at the level of the cavoatrial junction (preserving the sinus node and its artery), and the cardiac end is oversewn. At this point the superior vena cava is end-to-side anastomosed to the upper aspect of the right pulmonary artery, opened with a longitudinal or transverse incision, according to the surgical preference. This operation can be performed on normothermic cardiopulmonary bypass with a beating heart, therefore, avoiding aortic cross clamping and myocardial ischemia, and maintaining sinus rhythm. In certain circumstances, according to the surgical preferences, the cavopulmonary connection can be performed without cardiopulmonary bypass, using a temporary shunt or connection between the proximal superior vena cava or the innominate vein and the right atrium.

In patients with a persistent left superior vena cava, a bilateral bidirectional Glenn procedure (=end-to-side anastomosis, respectively, of the right superior vena cava to the right pulmonary artery and of the persistent left superior vena cava to the left pulmonary artery) is performed, with the same surgical technique for both the anastomoses.

Hemi-Fontan

The operation is performed on cardiopulmonary bypass with aortic cross clamping for its central portion. The medial aspect of the superior vena cava and the superior portion of the right atrium are incised as well as the confluence of the right and left pulmonary arteries. In the absence of pulmonary atresia, the proximal stump of the pulmonary artery is transected and oversewn. The superior vena cava is side-to-side anastomosed to the right pulmonary artery, and the same is done in the presence of a persistent left superior vena cava, which is side-to-side anastomosed to the ipsilateral pulmonary artery. A patch of biological (homograft, heterologous pericardium) or prosthetic (PTFE) material is used for augmentation of the pulmonary arteries anteriorly, for creation of a roof over the anastomosis of the vena cava and the pulmonary artery and a dam occluding the inflow of the superior vena cava into the right atrium, leaving a potential connection for the entire systemic venous return larger than the size of the inferior vena cava. An unrestrictive interatrial communication is also created, whenever necessary.

Modified Fontan

The completion of the Fontan circulation, deviating the systemic venous return from the inferior vena cava to the pulmonary arteries, can be accomplished with several different surgical techniques, according to the anatomy of the patient and the surgical experience.

- *Atriopulmonary connection*, consisting in a direct or indirect (with a roof of biological or synthetic material or with a conduit) connection between the right auricular appendage and either the transacted stump of the main pulmonary artery or the inferior aspect of the right pulmonary artery, associated with ventricular exclusion (Fig. 2.3.11), has now been practically abandoned because of the unfavorable fluidodynamics obtained with inclusion of the right atrium in this type of cavopulmonary connection.
- *Lateral tunnel* (Fig. 2.3.12 and 2.3.13) consists of the baffling with an interatrial patch (pericardium, PTFE) of the inferior vena cava to the superior vena cava and the upper portion of the right atrium, and then the superior vena cava and the upper portion of the right atrium are connected-with the inferior aspect of the right pul-

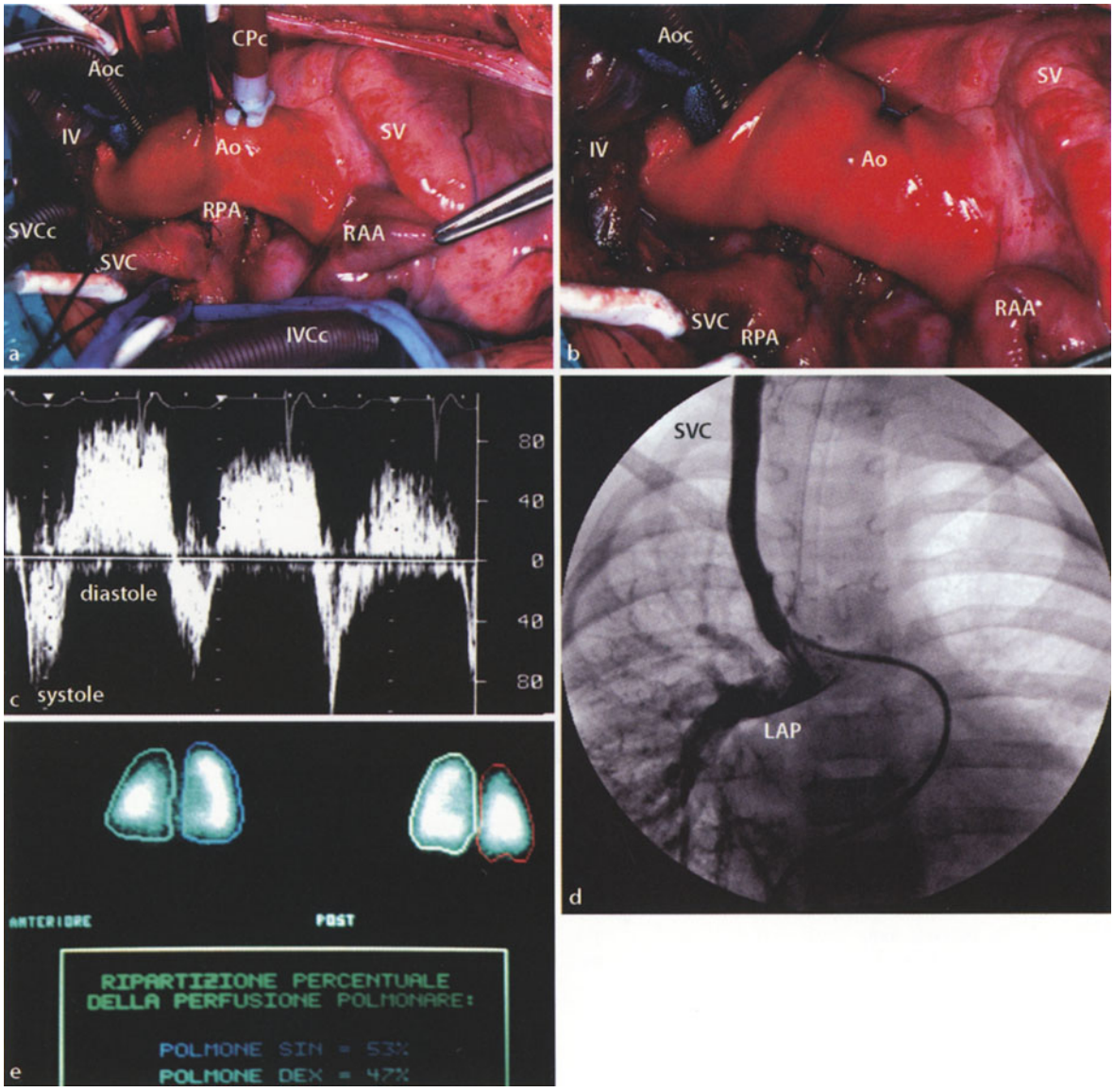

Fig. 2.3.10. Single ventricle: surgery. Bidirectional Glenn. **a** Intraoperative photograph of a bidirectional Glenn, with end-to-side anastomosis of the superior vena cava to the right pulmonary artery. The superior vena cava cannula is introduced directly into the innominate vein, while the two branches of the right pulmonary artery are controlled with the two elastic vessel loops. The cardioplegia cannula has been used in this specific case because of the need for an associated intracardiac procedure (*Ao* aorta, *Aoc* aortic cannula, *CPc* cardioplegia cannula, *IV* innominate vein, *IVCc* inferior vena cava cannula, *RAA* right auricular appendage, *RPA* right pulmonary artery, *SV* single ventricle, *SVC* superior vena cava, *SVCc* superior vena cava cannula), **b** intraoperative photograph of the same patient, after removal of all cannulas, with the exception of the aortic cannula, **c** postoperative transesophageal echocardiography showing the flow through the superior vena cava to the right pulmonary artery connection (bidirectional Glenn) with the superior vena cava-to-pulmonary artery flow during diastole and pulmonary artery-to-superior vena cava flow during systole, because of wash-out due to the antegrade pulmonary blood flow coming from the main pulmonary artery (photograph courtesy of Dr. Pierre-Guy Chassot), **d** postoperative angiography showing the injection in the superior vena cava with opacification of the right pulmonary artery; the contrast medium is diluted from the non-contrasted blood coming from antegrade pulmonary blood flow (*LPA* left pulmonary artery), **e** postoperative lung scintigraphy showing homogeneous distribution of the perfusion to both lungs, with 53% to the left lung and 47% to the right lung

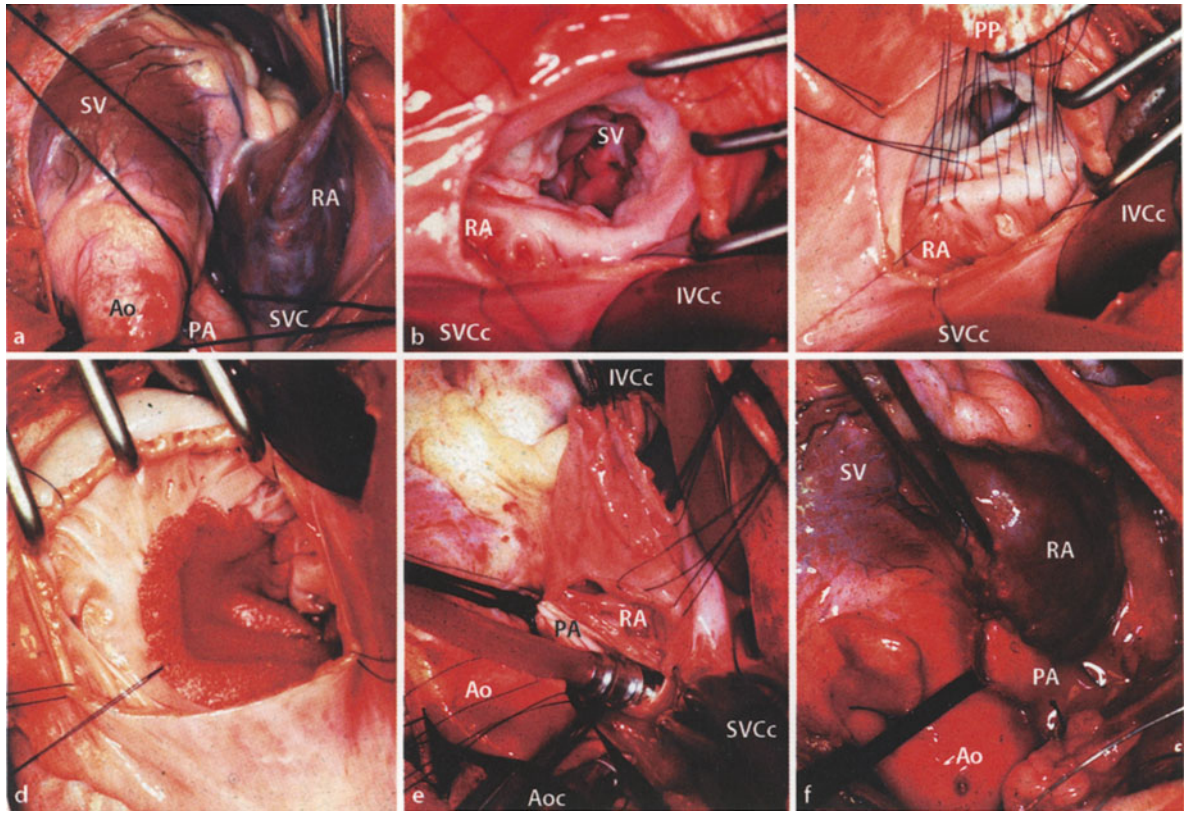

Fig. 2.3.11. Single ventricle: surgery. Modified Fontan with ventricular exclusion. Intraoperative photograph showing (**a**) the external anatomy of the single ventricle with ventriculoarterial discordance, and reduced pulmonary blood flow (*Ao* aorta, *PA* pulmonary artery, *RA* right atrium, *SV* single ventricle, *SVC* superior vena cava) and **b** through a right atriotomy, showing the inlet of the tricuspid valve into the single ventricle. (*IVCc* inferior vena cava cannula, *SVCc* superior vena cava cannula), **c** initial anastomosis of the prosthetic patch used for ventricular exclusion around the inlet of the tricuspid valve into the single ventricle (*PP* prosthetic patch), **d** completed anastomosis of the prosthetic patch used for ventricular exclusion around the inlet of the tricuspid valve into the single ventricle, initial (**e**) and completed (**f**) anastomosis of the opened right atrium to the inferior aspect of the right pulmonary artery, opened with a longitudinal incision, (*Aoc* aortic cannula, *IVCc* inferior vena cava cannula)

monary artery. The transected stump of the superior vena cava requires augmentation (by a roof of autologous or heterologous pericardium or PTFE) to accommodate the higher systemic venous return coming from the inferior vena cava. The coronary sinus is left in the lower pressure pulmonary venous atrium. This technique requires a period of aortic cross clamping with myocardial ischemia to open the right atrium and place the interatrial baffle. An unrestrictive interatrial communication is also created, whenever necessary.

Extracardiac connection (Fig. 2.3.14) consists of the extracardiac connection of the transected inferior vena cava (the cardiac stump is oversewn) to the inferior aspect of the right pulmonary artery by the interposition of a non-valved conduit (PTFE, pericardium, in situ pedicled pericardial tunnel), or in exceptional circumstances by direct anastomosis of the transected inferior vena cava to the transacted main pulmonary artery. The extracardiac connection can be performed on normothermic cardiopulmonary bypass with a beating heart, thereby avoiding aortic cross clamping and myocardial ischemia. In certain circumstances, according to the surgical preferences, the cavopulmonary connection can be performed even without cardiopulmonary bypass.

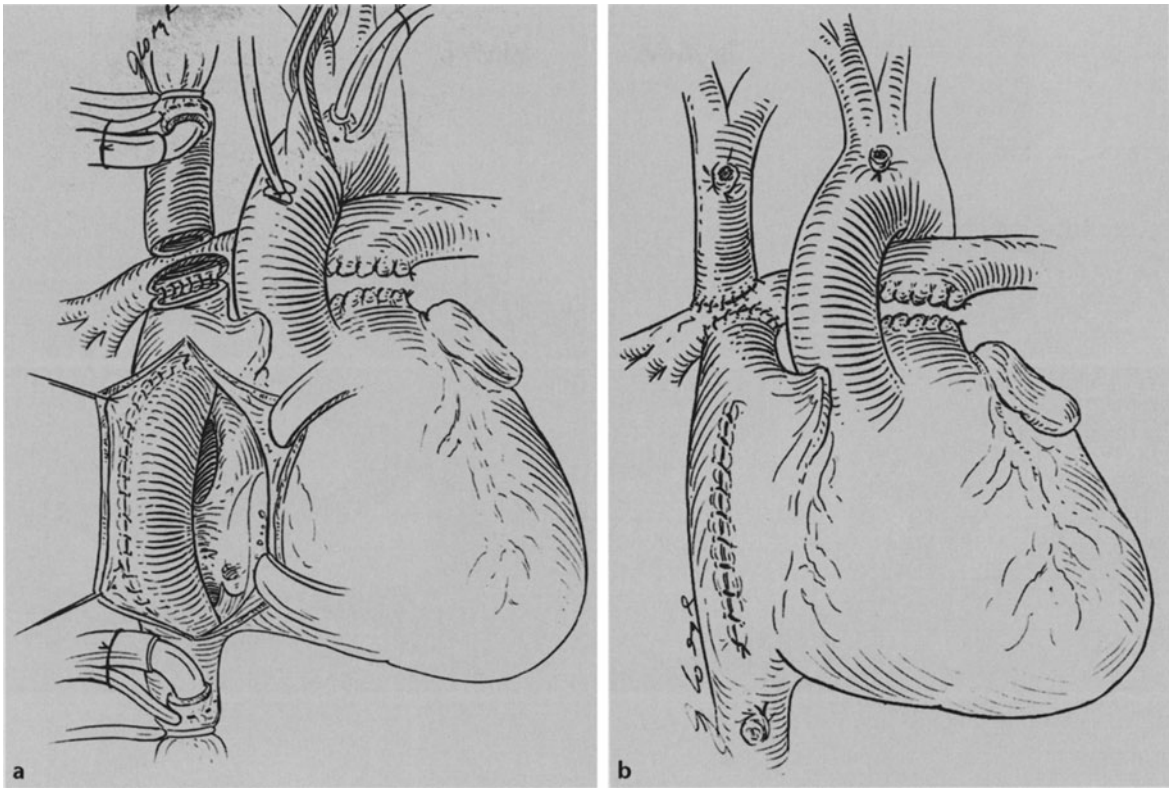

Fig. 2.3.12. Single ventricle: surgery. Modified Fontan: lateral tunnel. **a** Schematic drawing with the lateral tunnel technique of modified Fontan procedure, connecting the orifice of the inferior vena cava to the orifice of the superior vena cava with an interatrial PTFE patch sutured to the lateral wall of the right atrium (=lateral tunnel), **b** the total cavopulmonary connection with the lateral tunnel technique is completed with the end-to-side anastomosis of the transected superior vena cava to the superior aspect of the right pulmonary artery, while the atrial stump of the superior vena cava is end-to-side anastomosed to the inferior aspect of the right pulmonary artery (reproduced with permission from Laks H, Pearl JM, Haas GS, Drinkwater DC, Milgalter E, Jarmakani JM, Isabel-Jones J, George BL, Williams RG (1991) Partial Fontan: advantages of an adjustable interatrial communication. Ann Thorac Surg 52:1084–1094)

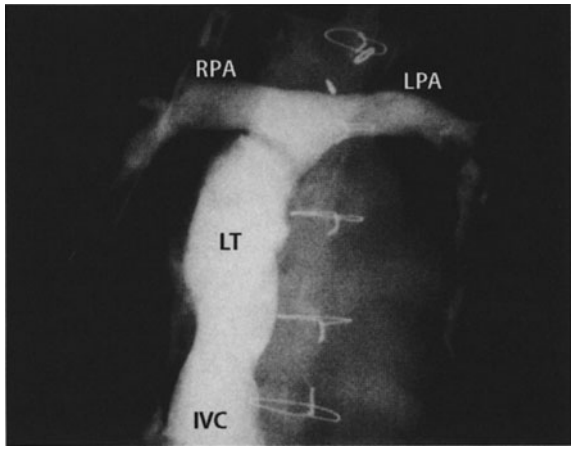

Fig. 2.3.13. Single ventricle: surgery. Modified Fontan: lateral tunnel. Postoperative angiography with contrast injection in the inferior vena cava, showing the lateral tunnel connecting the inferior vena cava with both pulmonary arteries (*IVC* inferior vena cava, *LPA* left pulmonary artery, *LT* lateral tunnel, *RPA* right pulmonary artery)

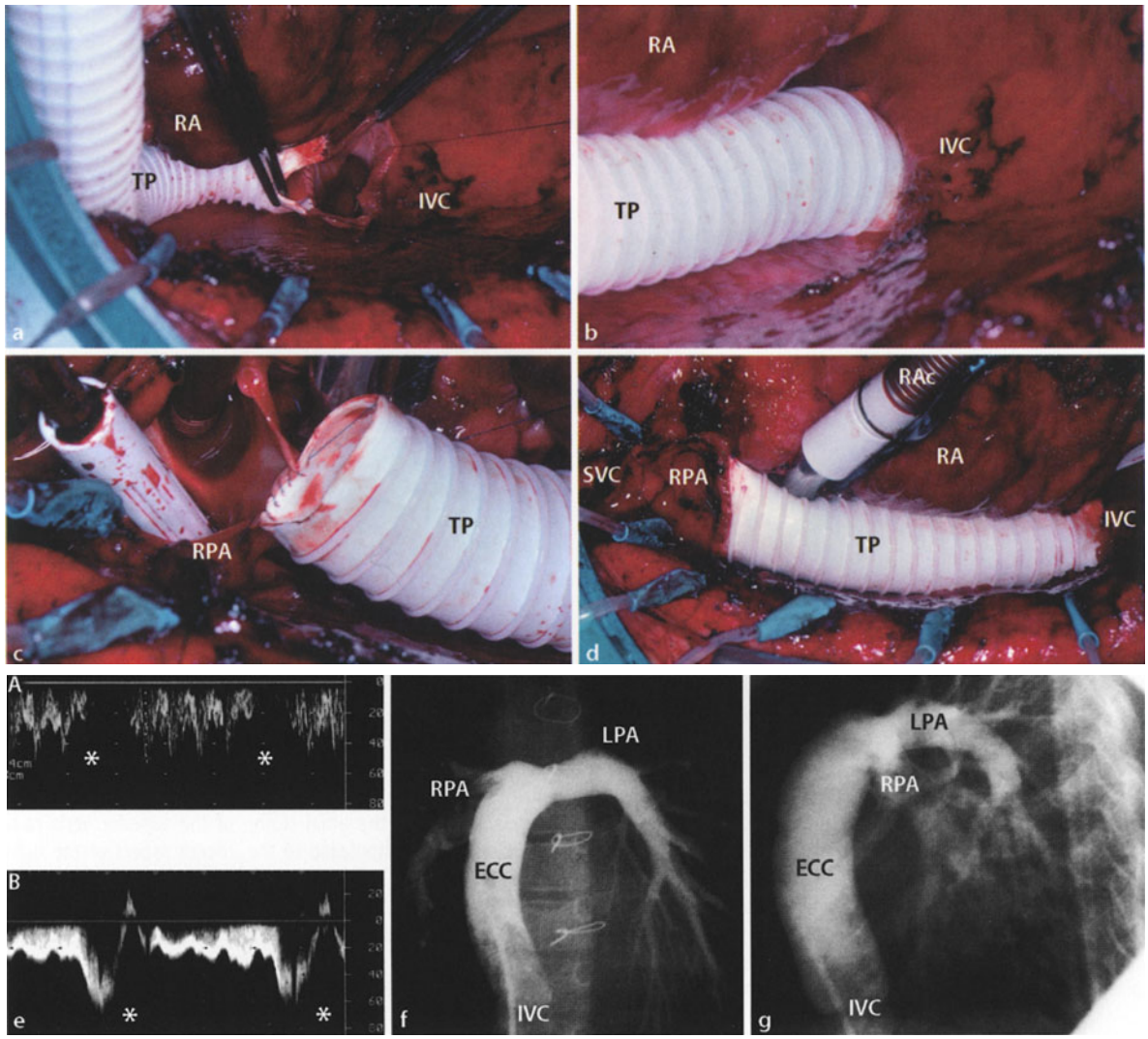

Fig. 2.3.14. Single ventricle: surgery. Extracardiac total cavopulmonary connection. **a** Proximal end-to-end anastomosis of a PTFE armed tubular prosthesis to the transected inferior vena cava; note the open technique, with the inferior vena cava unclamped and unsnared, and only a venous cannula introduced through the right femoral vein (*IVC* inferior vena cava, *RA* right atrium, *TP* tubular prosthesis), **b** completed proximal end-to-end anastomosis of the PTFE armed tubular prosthesis to the transected inferior vena cava, **c** distal end-to-end anastomosis of the PTFE armed tubular prosthesis to the longitudinally incision of the inferior aspect of the right pulmonary artery (*RPA* right pulmonary artery), **d** completed interposition of the PTFE armed tubular prosthesis between the inferior vena cava and the right pulmonary artery; note the previously performed end-to-side anastomosis between the superior vena cava and the superior aspect of the right pulmonary artery (bidirectional Glenn), and the right atrial cannula still in situ (*IVC* inferior vena cava, *RAc* right atrial cannula, *SVC* superior vena cava), **e** postoperative transesophageal echocardiography showing the flow through a total cavopulmonary connection with the different variations during inspirations (asterisks) related with intermittent positive pressure ventilation (**A**) and with spontaneous breathing (**B**) (photograph courtesy of Dr. Pierre-Guy Chassot), postoperative angiography in the anteroposterior view (**f**) and in the lateral view (**g**) showing the contrast medium injected in the extracardiac connection opacifying both pulmonary arteries. The contrast medium in the right pulmonary artery is diluted by the non-contrasted blood coming from the superior vena cava through the previously performed bidirectional Glenn (*ECC* extracardiac connection, *LPA* left pulmonary artery)

Fenestration (= incomplete atrial partitioning)

Different types of *fenestration* exist in order to allow for an incomplete separation between the caval and pulmonary venous pathways in order to decompress the systemic venous return. In the lateral tunnel technique, an interatrial communication (generally 4–5 mm diameter) is created in the central part of the prosthetic (PTFE) patch (= fixed fenestration), or is left on the lateral aspect of the suture of the patch, with a mattress suture outside the atrial wall to allow for an external adjustment (= adjustable atrial septal defect) by percutaneous manipulation of a snare.

In the extracardiac connection, the inferior vena cava to pulmonary artery conduit and the morphologically right atrium (now functioning as the pulmonary venous atrium = collecting chamber for the pulmonary veins and the coronary sinus) are off-bypass connected (= fenestration) either by a tubular prosthesis (PTFE, 6–8 mm diameter) end-to-side anastomosed between the extracardiac conduit and the morphologically right atrium or by a side-to-side direct anastomosis of the extracardiac conduit to the right atrium on two side-biting clamps. The fenestration obtained with a tubular prosthesis can also be percutaneously controlled with a snare.

All these types of fenestration will either spontaneously close in the late postoperative period, or they can be electively closed by a percutaneous intervention.

Ventricular septation

This procedure can be performed, on cardiopulmonary bypass, as a single stage, with a large patch dividing the two atrioventricular valves and the ventricular cavity, taking great care to avoid obstruction to the systemic and pulmonary outflow tracts. The size of the patch is crucial, since too large of a patch will bulge into the right ventricle in systole, impairing the right ventricular function; too small of a patch will result in an elevated incidence of dehiscence. The suturing lines of the patch will be dictated by the position of the tension apparatus of the two atrioventricular valves, the location of the two semilunar valves and the position of the ventricular septal defect.

Staged ventricular septation consists of placing an apical patch and a second patch at the superior portion between the atrioventricular valves, using widely spaced interrupted sutures, with the addition of a pulmonary artery banding; the ventricular septation (with debanding) is completed 6–18 months later with a third patch.

Potential complications

Pulmonary artery banding: potential for pulmonary artery distortion, development of pulmonary valvular regurgitation or damage to the pulmonary valve, progressive ventricular hypertrophy, decreased ventricular compliance, and development of subvalvular aortic stenosis. Subaortic stenosis often follows palliative pulmonary artery banding, in which the resultant hypertrophy of muscle around the ventricular septal defect may play a contributing role.

Bidirectional Glenn: prolonged pleural and/or pericardial effusions, chylothorax, phrenic nerve lesion. A persistently high pulmonary vascular resistance results in insufficient pulmonary blood flow and severe cyanosis, manifested by high superior vena cava pressure (over 18–20 mmHg). An elevated superior vena cava pressure may significantly decrease cerebral perfusion by decreasing the pressure gradient across the cerebral bed. This may be clinically manifested as fullness and pulsatility of the fontanelle, persistently irritability, systemic hypertension and relative bradycardia. If not reversible with conservative treatment by patient position with head elevated 45 degrees, early tracheal extubation to avoid positive pressure ventilation, aggressive pulmonary toilet, and pulmonary vasodilators (inhaled NO), the bidirectional Glenn may need to be taken down and replaced with a systemic-to-pulmonary artery shunt. Unusual following the bidirec-

tional Glenn procedure: the late development of pulmonary arteriovenous malformations or veno-venous collaterals may cause severe cyanosis. The latter result in right-to-left shunt, and, if severe, precludes successful conversion to Fontan operation.

Hemi-Fontan operation: distortion or degeneration of the biological or prosthetic materials used for the central pulmonary arteries augmentation and early or late supraventricular arrhythmias are potential complications in addition to those listed for the bidirectional Glenn procedure.

Modified Fontan procedure: in the presence a of failing Fontan procedure, a potential obstruction at any level of the cavopulmonary connections, as well as the persistency of intracardiac shunts (other than the surgically created fenestration), must be always ruled out. Rare cases have been reported with pulmonary venous pathway obstruction from restriction at the level of the atrial septum with the lateral tunnel technique.

The most frequent early complications are prolonged pleural effusion and chylothorax; chronic venous stasis (due to the high central venous pressure with low systemic vascular resistance, with subsequently increased hydrostatic capillary pressure), recurrent pericardial effusion, ascites, fluid retention, renal failure, hepatic failure and gastrointestinal dysfunction are more frequent in higher risk patients. While sinus node dysfunction, atrial fibrillation or flutter are more frequent, ventricular arrhythmias are less frequent, and complete atrioventricular block is very rare. Thromboembolism can occur in about 10% of patients, particularly in the presence of a low-output state with low velocity flow through the venous pathway; because of this risk these patients should be maintained under long-term prophylactic anti-platelets or anticoagulant treatment, according to policy of the hospital.

Protein-loosing enteropathy (reduced serum albumin concentration accompanied by diarrhea, poor appetite, failure to grow), pulmonary arteriovenous malformations and veno-venous collateral (between brachiocephalic angles and pericardial veins, azygos and hemiazygos system, Thebesian veins and epidiaphragmatic veins) formation with systemic arterial desaturation are the most frequent late complications.

Exercise intolerance (or reduced exercise capacity) is observed in a certain percentage of patients as well as progression of atrioventricular valve regurgitation.

Persistent and/or progressive hypoxemia can occur after the Fontan procedure, further increased with exercise, even in the absence of an evident intra-cardiac right-to-left shunt; the reason is probably the presence of a mild intrapulmonary shunt and the drainage of the coronary sinus into the pulmonary venous atrium.

Plastic bronchitis is very rare but mostly accompanied by dramatic consequences.

Either a fenestration (when absent or inadequate) or the early or late take-down of the total cavo-pulmonary connection is sometimes necessary, with or without associated treatment for the supraventricular arrhythmias. Sometimes becomes heart transplantation the only available solution, when the pulmonary vascular resistance is still within the normal range.

Ventricular septation: the incidence of complications with the ventricular septation, with the exception of very well selected cases, remains elevated. Potential complications are obstruction to the systemic and/or pulmonary inflow and/or outflow tracts, distortion of the atrioventricular valve(s) with resulting regurgitation, lesion of coronary arteries, residual interventricular shunt, complete atrioventricular block.

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Chapter 2.4 Pulmonary atresia with ventricular septal defect

Incidence

Pulmonary atresia with ventricular septal defect is a rare malformation, with an incidence of 0.042–0.070 per 1000 live births, or 1.5–3.4% of congenital heart defects, and a slightly higher prevalence in males than in females. Eighteen to 20% of patients with tetralogy of Fallot have pulmonary atresia.

Morphology (Figs. 2.4.1–2.4.3)

Hearts with pulmonary atresia with ventricular septal defect have a biventricular arrangement, concordant atrioventricular connections, unrestrictive ventricular septal defect, single aortic outlet and absent luminal continuity between the right ventricle and the pulmonary arterial circulation.

The ventricular septal defect, usually unrestrictive, is generally of the malalignment type, resulting from extreme anterior deviation of the infundibular septum, and extends nearly to the free wall of the right ventricle anteriorly; there might be other types of ventricular septal defect, occurring in any portion of the ventricular septum.

The aorta can be entirely connected to the left ventricle or may override the interventricular septum (26–50% of cases), with the aortic root, viewed from the ventricle, rotated clockwise.

Pulmonary atresia resulting from anterior deviation of the infundibular septum (like in tetralogy of Fallot) with underdevelopment of the right infundibular outflow tract and atretic infundibulum is most the common type (70% of the cases). More rarely pulmonary atresia can involve the pulmonary valve alone, with a thick fibrous membrane taking the place of the pulmonary valve, or with involvement of the pulmonary valve and the proximal portion of the main pulmonary artery, or it may involve a longer segment of the main pulmonary artery. In any case there is absence of luminal continuity between the right ventricle and the main pulmonary artery or both the right and left pulmonary arteries. The main pulmonary artery can be present and with reasonable size, but in most cases is severely hypoplastic; more rarely it consists of only a fibrous cord without lumen, and in 5% of the patients is completely absent.

The morphology of the central pulmonary arteries is highly variable. The central right and left pulmonary arteries may be present and communicate freely (confluent pulmonary arteries) or may not communicate (nonconfluent pulmonary arteries) or may be absent. Pulmonary artery discontinuity (nonconfluent pulmonary arteries) is present in 20–30% of patients. Stenosis at the origin of the right pulmonary artery is present in 10% of the cases, while there is stenosis of the left pulmonary artery in 20% of cases, probably because of the process of closure of the ductus arteriosus. The size of the pulmonary arteries at the prebranching site is extremely variable, and it can be quantitated with two different indexes: the McGoon ratio (see reference: Piehler JM), consisting of the ratio between the sum of the diameters of the right plus left pulmonary artery divided by the diameter of the descending thoracic

A. F. Corno, *Congenital Heart Defects*

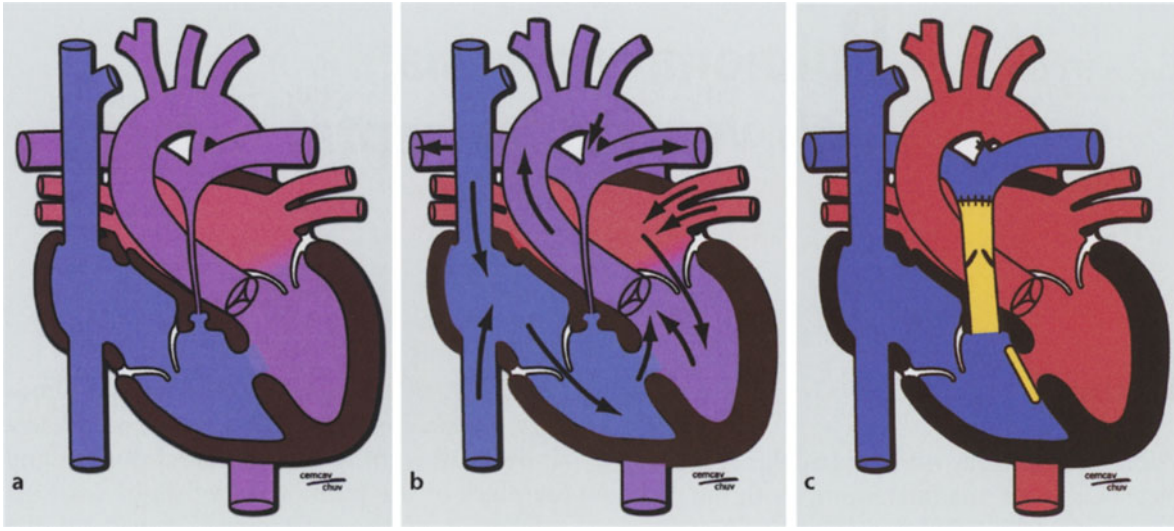


Fig. 2.4.1. Pulmonary atresia with ventricular septal defect, fibrous continuity between the right ventricle and pulmonary artery, and ductus-dependent pulmonary circulation: **a** morphology, **b** pathophysiology, **c** surgery

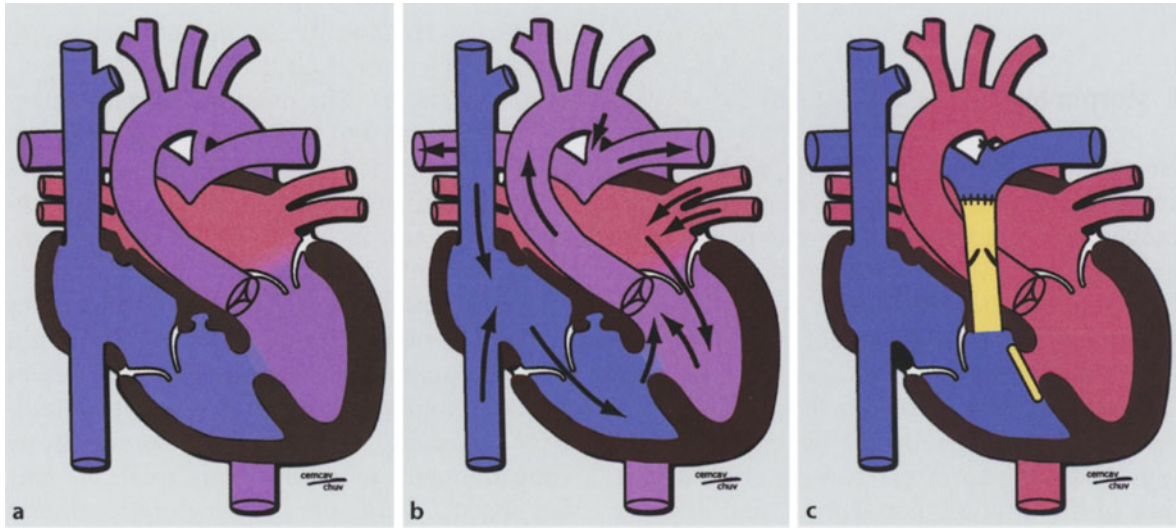


Fig. 2.4.2. Pulmonary atresia with ventricular septal defect, absence of continuity between the right ventricle and pulmonary artery, and ductus-dependent pulmonary circulation. **a** Morphology, **b** pathophysiology, **c** surgery

aorta at the level of the diaphragm, and the Nakata index (see reference: Nakata S), consisting of the ratio between the sum of the cross sectional areas of the right plus left pulmonary artery divided by the body surface area of the patient.

The pulmonary circulation presents with a high variability of situations (Fig. 2.4.4), and it may be supplied by a patent ductus arteriosus, by major aortopulmonary collateral arteries (present in at least 60–70% of cases), or by plexuses of bronchial and pleural arteries (5%).

The pathology of the intrapulmonary arteries depends on the source and amount of pulmonary blood flow. In the presence of a large patent ductus arteriosus supplying confluent pulmonary arteries, the blood flow and the intrapulmonary arteries of both lungs can be normal. In the absence of pat-

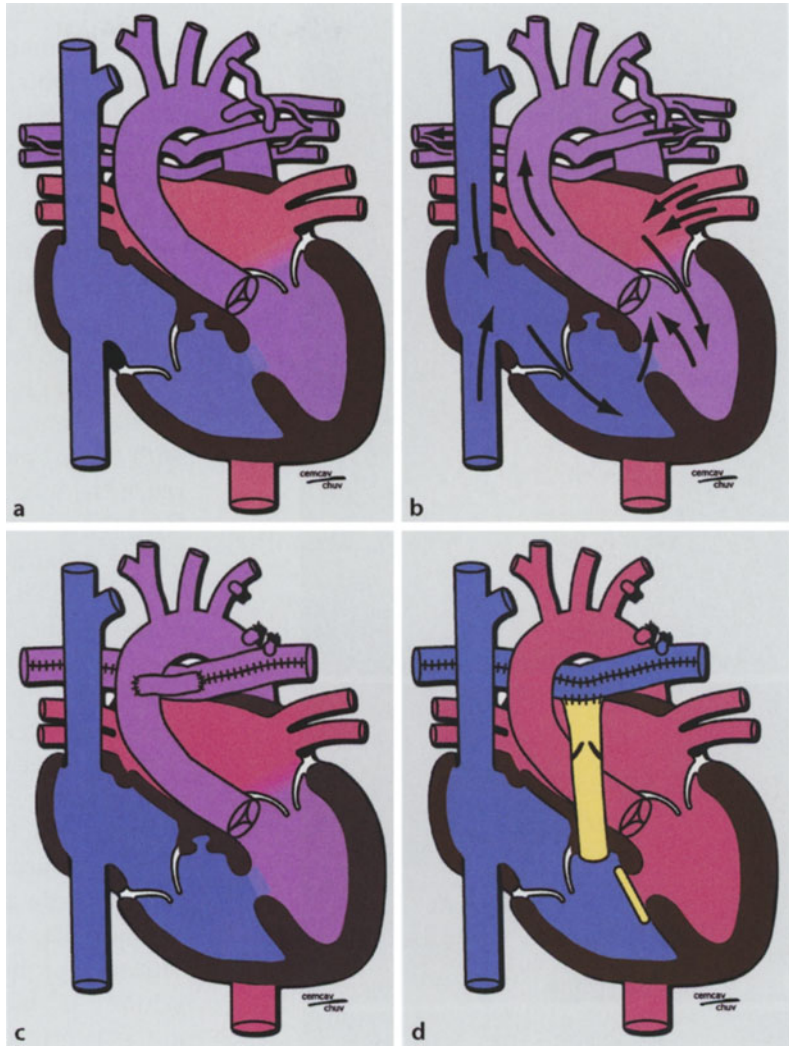
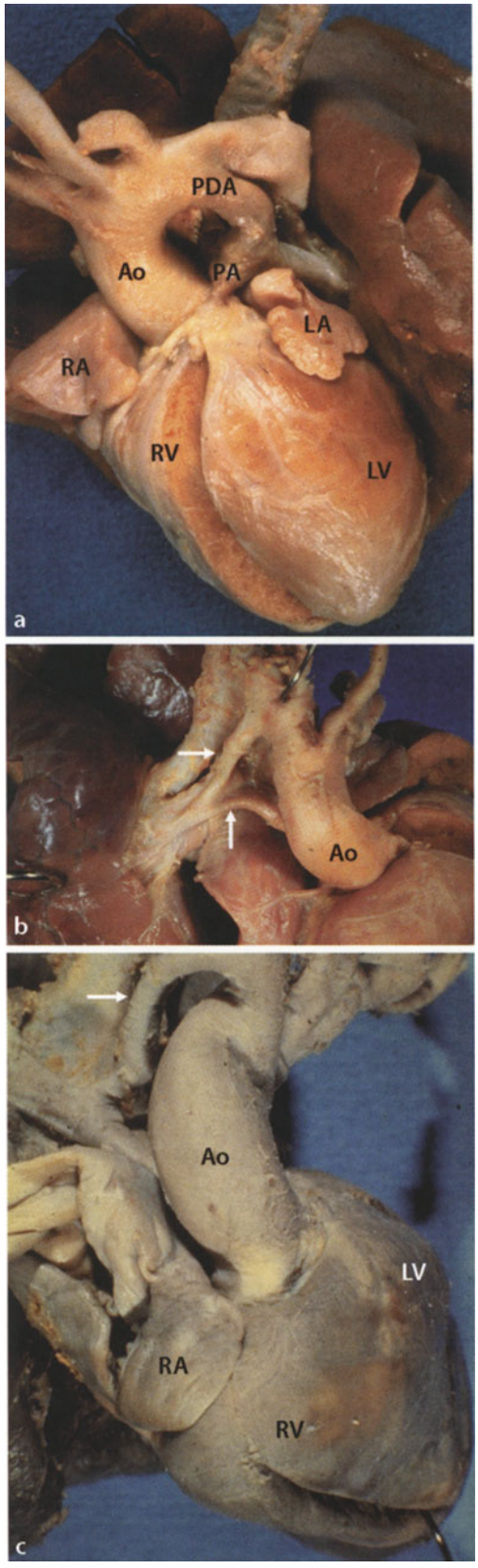


Fig. 2.4.3. Pulmonary atresia with ventricular septal defect and major aortapulmonary collateral arteries. **a** Morphology, **b** pathophysiology and surgery, **c** palliation: unifocalization and central shunt, and **d** repair

ent ductus arteriosus and the presence of multiple major aortopulmonary collateral arteries, the consequence is an abnormal intrapulmonary arborization, with dishomogeneous morphology of the intrapulmonary arteries among different pulmonary segments. Only about half (53%) of patients with confluent pulmonary arteries present with pulmonary arteries reaching all 20 pulmonary segments, while less than 20% of patients with nonconfluent and/or hypoplastic pulmonary arteries have complete distribution of pulmonary arteries to all pulmonary segments. The major aortopulmonary collateral arteries may anastomose at any site in the pulmonary vascular tree: extrapulmonary, hilar, lobar and segmental level. Rare cases have been reported with mixed source of pulmonary blood flow, with the simultaneous presence of patent ductus arteriosus and major aortopulmonary collateral arteries. In these cases the nature and distribution of the pulmonary arterial supply is often very complex. Exceptional reports exist with the presence of bilateral (right and left) ductus arteriosus.

The orientation and position of the ductus arteriosus, when present, are abnormal, with a downward direction from beneath the left aortic arch; also size and shape are quite variable, while in most patients the ductus is longer and more tortuous than usual.



Major collateral arteries are systemic arteries originating from the splanchnic arterial network which persist after birth and provide blood to the lungs by different modalities. They arise typically from the descending thoracic aorta, less commonly from the subclavian arteries, and rarely from the internal mammary (thoracic) arteries, intercostal arteries, carotid arteries or abdominal aorta; extremely rare cases have been reported with collateral arteries arising from coronary arteries. After a complex course in the posterior mediastinum where relationships are taken with the esophagus and the airway, collaterals reach the lungs and provide pulmonary blood flow either terminally or communicating with native proximal or distal pulmonary arterial branches. They may feed one side of the lungs, both sides or cross to the opposite side. Non-communicating collaterals branching within the pulmonary parenchyma have the same histologic characteristics of normal pulmonary arteries and may undergo hypertensive vascular changes exactly as normal pulmonary vasculature does. Because of processes of intimal proliferation, major collaterals can develop stenosis in up to 60% of the patients. Stenoses are thought to be a consequence of the shear stress related to the high velocity of blood flow due to the aortapulmonary pressure gradient. They may evolve quite variably and unpredictably, and may have a protective effect on pulmonary microvasculature. However, they may also complicate the surgical treatment of the disease.

Fig. 2.4.4. Pulmonary atresia with ventricular septal defect: morphology. **a** Anatomical fibrous continuity between the right ventricle and the main pulmonary artery, perfused by a patent ductus arteriosus. (*Ao* aorta, *LA* left atrium, *LV* left ventricle, *PA* pulmonary artery, *PDA* patent ductus arteriosus, *RA* right atrium, *RV* right ventricle) (reproduced with permission from Marino B, Thiene G (1990) Atlante di anatomia ecocardiografica delle cardiopatie congenite, USES, Firenze, **b** major aortopulmonary collateral arteries (white arrows) originating from the aorta and its branching and perfusing the lung (photograph courtesy of Dr. Bruno Marino), **c** major aortopulmonary collateral artery (white arrow) orignating from the aorta, which branches and perfuses the lung (photograph courtesy of Dr. Bruno Marino)

Associated anomalies

Like tetralogy of Fallot with pulmonary stenosis, 25% of these patients have a dilated right aortic arch, although some estimate that is thought to be as frequent as 50%. Other anomalies identified in association with pulmonary atresia with ventricular septal defect include dextrocardia, heterotaxia, anomalous pulmonary or systemic venous connections, atrial septal defect or patent foramen ovale (50%), atrioventricular septal defect, tricuspid atresia, double inlet single left ventricle, aortic valve stenosis, complete transposition of the great arteries, double discordance (=congenitally corrected transposition of the great arteries), anomalous coronary arteries like in tetralogy of Fallot or coronary artery-to-pulmonary artery fistulas, reported in about 10% of patients with pulmonary atresia with ventricular septal defect, from either the left (most frequently) or the right coronary artery, in all cases joining the central pulmonary artery (see chapter "Anomalous coronary arteries").

Pulmonary atresia with ventricular septal defect is one of the most common cardiac defects associated with various syndromes: DiGeorge (=velocardiofacial syndrome with deletion of chromosome 22q11.2, previously called CATCH-22=cardiac defect, abnormal face, thymic hypoplasia, cleft palate, hypocalcemia, microdeletion of band 22q11), VATER (=vertebral defects, anal atresia, tracheoesophageal fistula with esophageal atresia, renal and radial anomalies), CHARGE (=coloboma, heart disease, atresia choanae, retarded growth, retarded development and/or central nervous system anomalies, genital hypoplasia, ear anomalies and/or deafness), Alagille, cat's-eye, de Lange, Klippel-Feil, Down. Patients with deletion of chromosome 22q11.2 present a higher incidence of hypoplastic pulmonary arteries (41%) than presence of confluent pulmonary arteries (16%).

Pathophysiology

Pulmonary atresia with ventricular septal defect demonstrates a very wide spectrum of pathophysiologic patterns and severity, from elevated pulmonary blood flow through patent ductus and collaterals to very reduced pulmonary blood flow. The source, volume and distribution of the pulmonary arterial supply is often very complex. In the majority of patients (60%) the pulmonary arterial pressure tends to be normal or below normal because of the presence of stenoses of the collateral arteries either at the proximal (systemic) origin or at intrapulmonary sites, and because the stenoses tend to progress over time in untreated children. Pulmonary hypertension is not frequently encountered in these patients, but it may be present or develop in lung segments perfused by unrestricted arterial supply.

Diagnosis

Clinical pattern:

- the age at presentation may vary depending on the total amount of pulmonary blood flow; the great majority of patients come to observation in the neonatal period with cyanosis because of the closure of the ductus arteriosus; when the ductus arteriosus is the only source of pulmonary blood flow, cyanosis may be very severe; patients with reduced pulmonary blood flow coming later to observation present with severe cyanosis and polycytemia; in the presence of large patent ductus arteriosus or multiple collateral arteries, well developed, presentation may be delayed and the symptom cyanosis is not very clear, since there is an increased pulmonary blood flow and the patients present with congestive heart failure more than cyanosis; patients may also present later with progressive cyanosis because growth outstrips the pulmonary blood flow; survival through adulthood has been described in a few patients with well-devel-

oped collateral arteries; respirators distress with persistent airway hyperresponsiveness is one of the most common problems, because of the tracheobronchomalacia due to the external airways compression by the hypertensive collaterals, particularly with the association of the right aortic arch; hemoptysis can occur as the result of rupture of dilated systemic-pulmonary collateral arteries; on auscultation, the second cardiac sound is always single and often accentuated, a systolic murmur is audible along the lower left sternal border, and a continuous murmur is heard over the upper chest in the presence of patent ductus arteriosus, while it may diffusely audible over the entire chest and in the back in the presence of major aorta-pulmonary collateral arteries.

Electrocardiogram:
- right axis deviation with right atrial and right ventricular hypertrophy.

Chest X-ray:
- classical imaging of boot-shaped heart, due to upturned heart caused by the right ventricular hypertrophy and the concavity in the region of the main pulmonary artery produced by the underdevelopment of the entire right ventricular outflow tract (Fig. 2.4.5); the aorta is generally dilated, with a right aortic arch in 25–50% of patients.

Echocardiogram:
- the parasternal long-axis view shows a large aortic valve overriding a malalignment ventricular septal defect, and is also used to detect anomalous coronary arteries; the parasternal cross sectional view confirms the presence of a blind hypoplastic right ventricular outflow tract; the suprasternal and high parasternal views provide information on presence, confluence and size of the pulmonary arteries, patency of the ductus arteriosus and side of the aortic arch; color flow imaging identifies the sources of pulmonary blood flow, including patent ductus arteriosus and/or major aortopulmonary collaterals.

Cardiac catheterization:
- cardiac catheterization is necessary in all patients, with the exception of those severely cyanotic and requiring an urgent systemic to pulmonary artery shunt because of ductus-dependent pulmonary circulation;
- in the cardiac catheterization the presence, confluence, size and distribution of the true pulmonary arteries should be identified as well as the collateral circulation (Figs. 2.4.6–2.4.10), the coronary artery anatomy, and the presence of any other ventricular septal defect that might be associated with a ventricular malalignment septal defect;

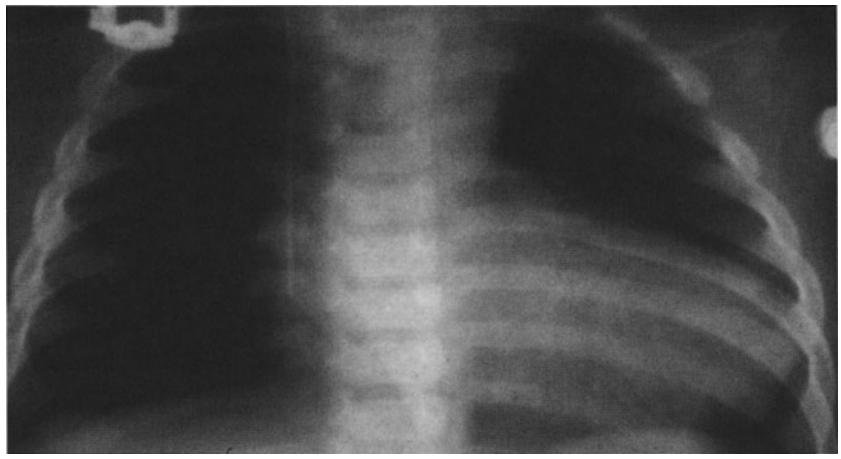


Fig. 2.4.5. Pulmonary atresia with ventricular septal defect: radiology. Anteroposterior chest X-ray with the classical imaging of boot-shaped heart, with poor lung perfusion

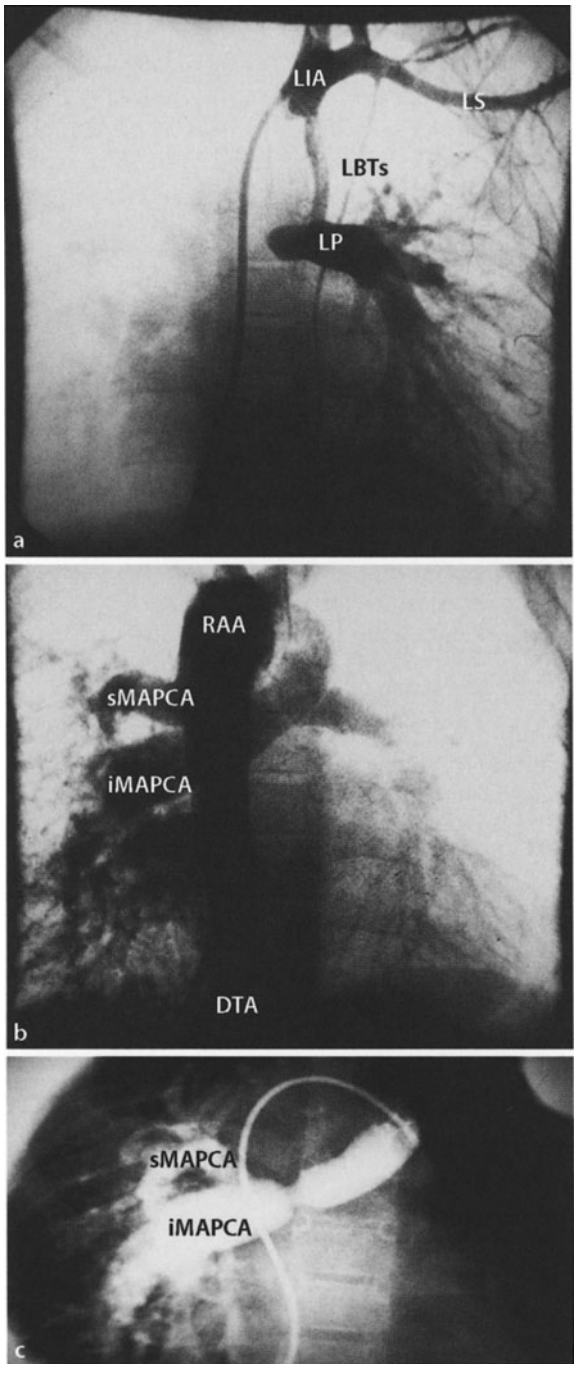


Fig. 2.4.6. Pulmonary atresia with ventricular septal defect: angiography. **a** Anteroposterior view with contrast injection in the left-sided innominate artery (because of the presence of the aortic arch) showing the left pulmonary artery opacified from a previously modified left Blalock-Taussig shunt in a patient with pulmonary atresia, ventricular septal defect, disconnected pulmonary arteries, right aortic arch (*LBTs* left modified Blalock-Taussig shunt, *LIA* left-sided innominate artery, *LPA* left pulmonary artery, *LSA* left subclavian artery), **b** contrast injection in the same patient showing the right aortic arch giving origin to a hypertensive (systemic pressure) major aortopulmonary collateral artery perfusing the middle and inferior right lobes, and the descending thoracic aorta giving origin to a hypertensive (systemic pressure) major aortopulmonary collateral artery perfusing the right upper lobe; there is confirmation that the pulmonary arteries are disconnected (*DTAo* descending thoracic aorta, *iMAPCA* inferior major aortapulmonary collateral artery, *RAA* right aortic arch, *sMAPCA* superior major aortopulmonary collateral artery), **c** contrast injection in the same patient showing the major aortopulmonary collateral artery perfusing the right upper lobe after surgical disconnection from the aorta and end-to-side anastomosis to the other major aortopulmonary collateral artery perfusing the middle and inferior right lobes, and banding (black arrow) of the proximal segment of the second major aortopulmonary collateral artery, reducing the distal pressure to half the systemic value; the patient subsequently underwent successful repair with closure of the ventricular septal defect, pericardial roll reconstruction of the pulmonary arteries confluence and biological valved conduit implantation between the right ventricle and the new pulmonary arteries bifurcation

- hypoplastic confluent pulmonary arteries are visualized on angiography with a characteristic "sea-gull" aspect on the anteroposterior view; they can be shown by retrograde filling from a collateral injection or a pulmonary venous wedge injection and by direct filling via a collateral or a previous shunt; in the presence of collaterals, they should be first investigated by a thoracic descending aortogram, followed by selective pressure measurement and angiography, possibly with a balloon inflated for temporary distal aortic occlusion;
- both collaterals and native pulmonary arteries (when present) should be classified for size, number of perfused lung segments, and areas of dual pulmonary blood supply, the calculation of indexed cross-sectional areas of pulmonary arteries (*pulmonary artery index*) and of collaterals (*total collateral index*), as well as their sum (*total neopulmonary artery index*), may be helpful for surgical planning;
- patients with increased pulmonary blood flow and congestive heart failure could benefit from coil occlusion of collateral arteries;

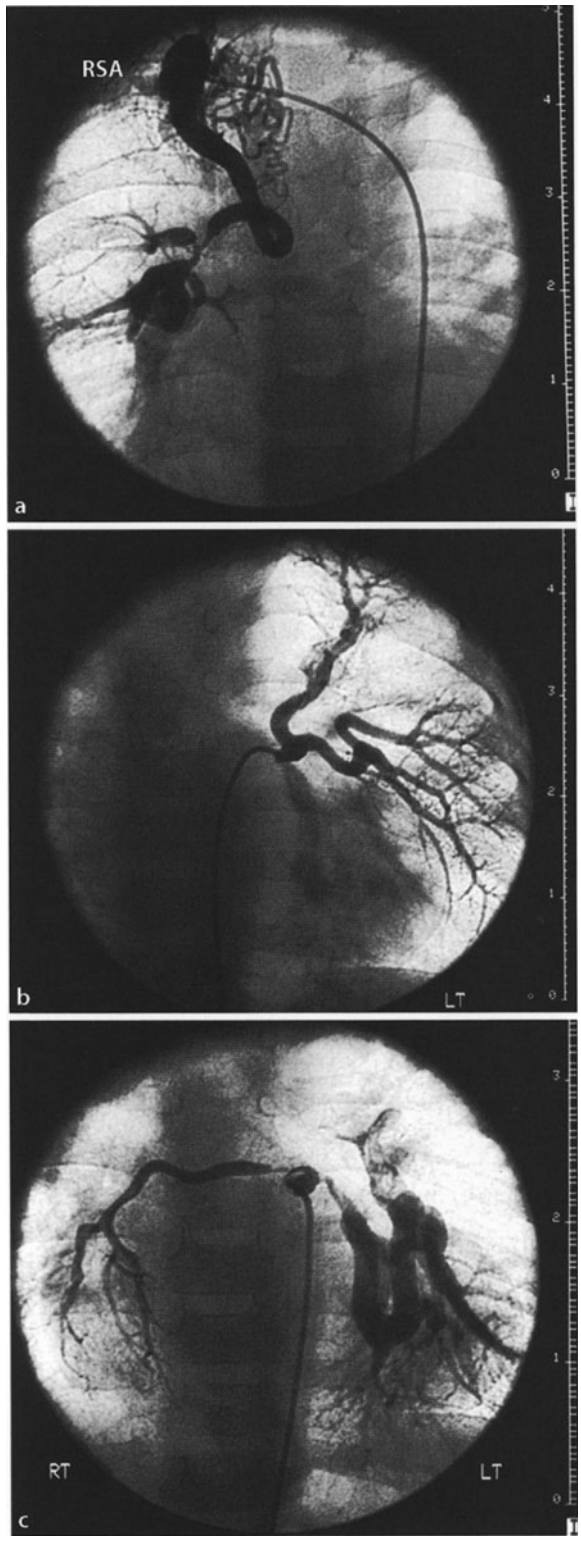


Fig. 2.4.7. Pulmonary atresia with ventricular septal defect: angiography. **a** Anteroposterior view with contrast injection at the origin of the right subclavian artery in a child with pulmonary atresia, ventricular septal defect and major aorto pulmonary collateral arteries, showing the collateral arteries perfusing the right lung (*RSA* right subclavian artery), **b** angiocardiography of the same patient with contrast injection from the descending thoracic aorta at the origin of collateral arteries perfusing the left lung, **c** angiocardiography of the same patient with contrast injection from the descending thoracic aorta at the origin of multiple collateral arteries perfusing the right and the left lung

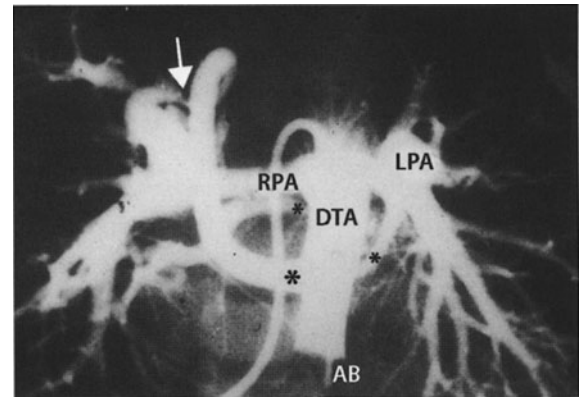


Fig. 2.4.8. Pulmonary atresia with ventricular septal defect: angiography. Angiocardiography with balloon occlusion of the distal descending thoracic aorta and contrast injection showing the origin of multiple collateral arteries (black asterisk), one with an evident stenosis (white arrow), perfusing the right and the left lung (*AB* aortic balloon, *DTAo* descending thoracic aorta, *LPA* left pulmonary artery, *RPA* right pulmonary artery

- pulmonary artery pressure and resistance should be evaluated in older patients with unrestrictive communications between the systemic and the pulmonary circulations to rule out pulmonary vascular obstructive disease.

Indications for surgical treatment

Identification of the systemic pulmonary blood supply is essential in planning the type of surgical approach. In neonates with ductus-dependent pulmonary blood flow, prostaglandin administration is often required to keep the ductus arteriosus open until surgery can be performed. Various surgical options are available, depending upon the heterogeneity of the anatomy and pathophysiology of the individual patient, especially in the presence of collateral arteries.

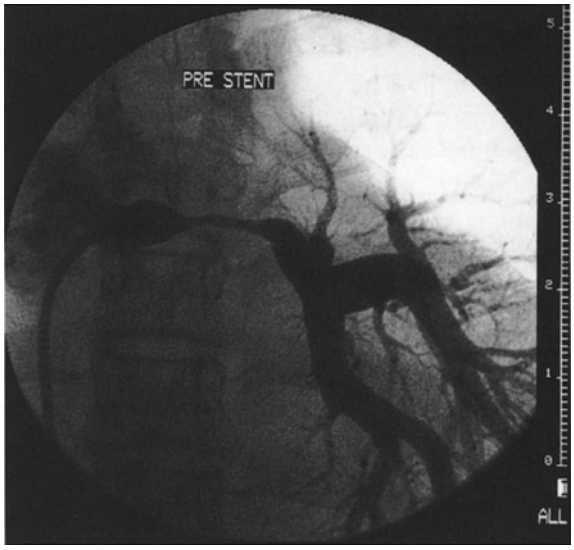


Fig. 2.4.9. Pulmonary atresia with ventricular septal defect: angiography. Anteroposterior view with contrast injection showing a stenotic segment of a major aortopulmonary collateral artery immediately after its origin from the descending thoracic aorta, and before its branching for the left lung; this morphology is suitable for dilatation and stent implantation to recruit the perfusion to the left lung

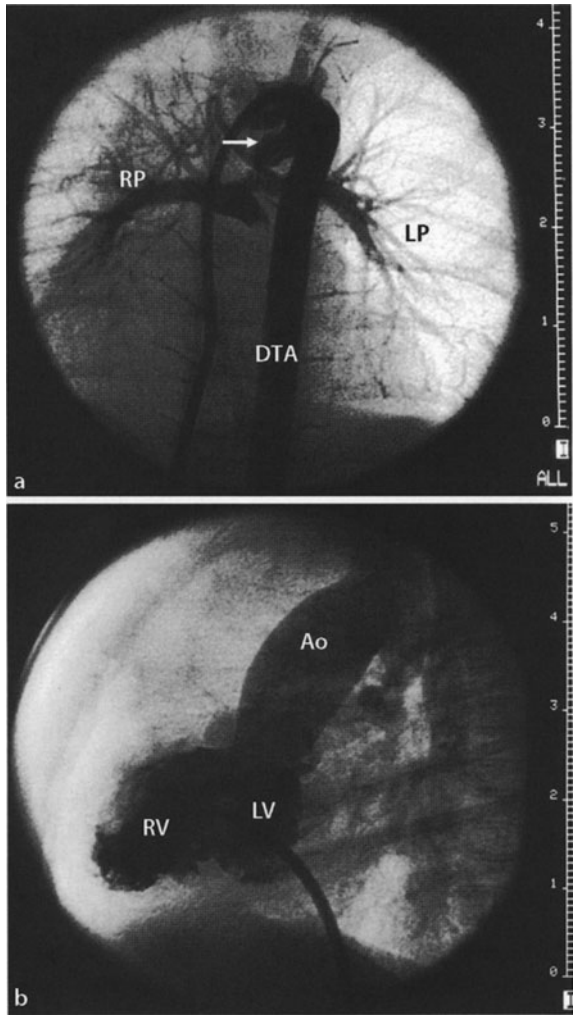


Fig. 2.4.10. Pulmonary atresia with ventricular septal defect: angiography. **a** Anteroposterior view in a neonate with pulmonary atresia, ventricular septal defect, confluent pulmonary arteries and ductus-dependent pulmonary blood flow, with contrast injection in the aorta near the origin of the patent ductus arteriosus (white arrow); this morphology is favorable for surgical treatment (*DTAo* descending thoracic aorta, *LPA* left pulmonary artery, *RPA* right pulmonary artery), **b** lateral view in the same neonate with contrast injection in the right ventricle and opacification of the left ventricle and aorta, single outlet for the heart (*Ao* aorta, *LV* left ventricle, *RV* right ventricle)

In order to achieve the ultimate goal of a complete repair in such complex situation:

- First: multifocal pulmonary blood supply needs to be converted to a single source through so – called "unifocalization" procedures;
- Second: the neopulmonary vascular bed must be large enough to receive the whole cardiac output;
- Third: the pulmonary vascular tree needs to be connected to as many as possible undamaged lung segments.

Three-quarters of the total lung capacity, or 15 of the 20 lung segments or 1½; lung should be incorporated in the unifocalization in order to obtain the best functional results.

Palliation

In the presence of hypoplastic, nonconfluent or absent pulmonary arteries, or inadequate peripheral arborization, only a palliative approach can be take into consideration as the initial surgical treatment.

- *Modifiefd Blalock-Taussig shunt:* the most used palliative surgical approach to increase the pulmonary blood flow in cyanotic patients, nowadays finds a very limited application in children with pulmonary atresia with ventricular septal defect, because of frequent reports of distortion with or without acquired discontinuity of the pulmonary arteries, and occasionally

development of pulmonary hypertension in the supplied lung segments.

- *Palliative right ventricular outflow tract reconstruction:* growth of pulmonary arteries, enabling later complete repair, can be obtained by increasing their perfusion pressure and flow through a right ventricular outflow tract reconstruction. If collateral arteries exist and provide terminal perfusion to the otherwise excluded lung segments, then incorporation of those collateral arteries through a process of unifocalization should be considered. Collateral arteries responsible for dual perfusion can be surgically ligated or coil-occluded with percutaneous intervention.
- *Creation of an aortopulmonary window:* In patients considered as unsuitable candidates for single-stage unifocalization and complete repair because of the presence of poorly developed aortopulmonary collateral arteries and hypoplastic pulmonary arteries, a recently introduced surgical option is the creation of an aortopulmonary window. The selection criteria include: 1) presence of centrally confluent true pulmonary arteries 1.0 to 2.5 mm in diameter, with a well-developed peripheral arborization pattern, 2) multiple small aortopulmonary collaterals, most of which are in communication with the true pulmonary arterial system; 3) severe cyanosis. This surgical approach can increase the size of the true pulmonary arteries, making these patients better candidates for eventual complete repair.
- *Unifocalization with central shunt or right ventricular outflow tract reconstruction without closure of the ventricular septal defect:* in the presence of bilateral major aortopulmonary collateral arteries with different origin of the pulmonary blood source, either sequential unilateral unifocalization (more rarely in infants and more frequent in older patients with late referral) or simultaneous bilateral unifocalization is performed, with either a central shunt (PTFE tubular prosthesis connecting the ascending aorta to the reconstructed pulmonary artery confluence) or a valveless conduit (PTFE or pericardial) or a biological valved conduit interposed between the right ventricle and the pulmonary artery reconstructed confluence (leaving the ventricular septal defect open) to provide unrestrictive pulmonary blood flow and to homogeneous development of the pulmonary arteries.
- *Reduction of pulmonary blood flow and pressure:* major aortopulmonary collateral arteries with excessive pulmonary artery blood flow and pressure, when they cannot be either included in the unifocalization process or occluded by coil in the catheterization laboratory, need to be interrupted where there is an alternative source of blood flow to the lung segments involved, or a banding of these collateral arteries has to be considered, particularly in older children in the presence of collateral arteries with systemic pressure.

Repair

Complete repair consists of complete separation of the pulmonary and systemic circulations by closure of the atrial and ventricular septal defects, closure of all extracardiac sources of pulmonary blood flow, and establishment of unobstructed continuity between the right ventricle and pulmonary arteries (or the reconstructed confluence of the bilateral pulmonary blood supply) with interposition of an extracardiac valved conduit.

Single stage unifocalization of pulmonary blood supplies and complete intracardiac repair is the procedure of choice in infants, leaving the single stage unifocalization with postponed closure of the ventricular septal defect as an alternative surgical approach in the absence of the criteria for single stage complete repair.

The criteria for complete surgical repair are the following:

- in the presence of intrapericardial pulmonary arteries, their cross-sectional area must be more than 50% of the normal value for the age and body surface of the patient;

- the pulmonary arteries must supply at least 10 lung segments, or the equivalent of one lung;
- in the presence of a single pulmonary artery, it must have normal size and supply all segments of that lung.

Heart-lung transplantation.

This is an extreme surgical option to be considered for end-stage patients unsuitable for any alternative surgical treatment or with demonstrated diffused pulmonary vascular obstructive disease.

Individualized integrated approach.

An "*individualized integrated approach*" was proposed by Adriano Carotti (see references), and has been further developed since, in order to increase the probability of reaching complete surgical repair, regardless of the anatomical complexity. According to the "*individualized integrated approach*", patients undergo a preoperative selection mainly based on the indexed measurements of the pulmonary blood flow sources. The main criterion is the preoperative calculation of the *total neopulmonary arterial index* (Fig. 2.4.11), resulting from the combined cross-sectional area of both native pulmonary arteries and major aortopulmonary collateral arteries indexed to the body surface area. A value of *total neopulmonary arterial index* equal or greater than 150 mm^2/m^2 is indicative of an overall compliance of the pulmonary vascular tree adequate to accommodate single-stage unifocalization; these patients can undergo through median sternotomy a complete unifocalization on cardiopulmonary bypass, and the suitability for closure of the ventricular septal defect is assessed intraoperatively by a pulmonary blood flow-study. Patients with a *total neopulmonary arterial index* less than 150 mm^2/m^2 are primarily treated by a palliative surgical approach aimed at increasing the forward blood flow into the true pulmonary arteries to promote their growth, to possibly allow subsequent midline unifocalization and later complete repair.

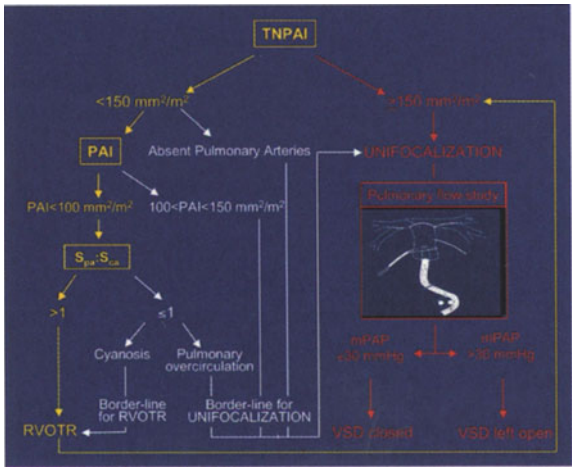


Fig. 2.4.11. Pulmonary atresia with ventricular septal defect: indication for surgery. Individualized integrated approach, based on the measurement of the total neopulmonary arterial index (*mPAP* mean pulmonary artery pressure, *PAI* pulmonary arterial index, *RVOTR* right ventricular outflow tract reconstruction, *TNPAI* total neopulmonary arterial index, *VSD* ventricular septal defect) (reproduced with permission from Carotti A, Albanese SB, Minniti G, Guccione P, di Donato RM (2003) Increasing experience with integrated approach to pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries. Eur J Cardiothorac Surg 23:719–727)

Alternative surgical approach

In the rare presence of major aortopulmonary collaterals all originating from a relatively short segment of the descending thoracic aorta, a possibility is to perform a single stage repair using the transected segment of the descending thoracic aorta, containing the origin of collaterals, connected to the right ventricle via a biological valved conduit.

Surgical treatment

Creation of an aortopulmonary window

Through a median sternotomy, without cardiopulmonary bypass, the hypoplastic confluence of the true pulmonary arteries is dissected free and then end-to-side anastomosed to the lateral aspect of the ascending aorta, by means of a temporary side aortic clamp (Fig. 2.4.12)

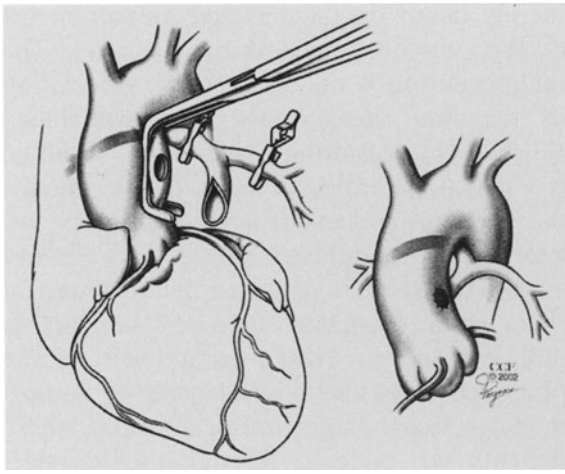


Fig. 2.4.12. Pulmonary atresia with ventricular septal defect: surgery. Palliation: central end-to-side shunt (=Melbourne shunt) (reproduced with permission from: Duncan BW, Mee RBB, Prieto LR, Rosenthal GL, Mesia CI, Qureshi A, Tucker OP, Rhodes JF, Latson LA (2003) Staged repair of tetralogy of Fallot with pulmonary atresia and major aortopulmonary collateral arteries. J Thorac Cardiovasc Surg 126:694–702)

Unifocalization with central shunt or right ventricular outflow tract reconstruction without closure of the ventricular septal defect

(Figs. 2.4.13–2.4.15)

Through a median sternotomy all the major aortapulmonary collateral arteries are identified through the transverse sinus and posterior mediastinal dissection, controlled and snared just before the beginning of cardiopulmonary bypass. Collateral arteries are then separated from their systemic origin, mobilized toward the posterior mediastinum and joined together to construct a pulmonary artery confluence, either with direct anastomoses or with the interposition of an autologous or heterologous pericardial roll (Figs. 2.4.16 and 2.4.17); the pulmonary blood flow is obtained either with a central shunt (PTFE tubular prosthesis proximally anastomosed end-to-side to the lateral aspect of the ascending aorta and distally end to side anastomosed to the reconstructed pulmonary artery confluence) or a valveless conduit (PTFE or pericardial) (Figs. 2.4.18), or a biological valved conduit interposed between a right ventriculotomy and the pulmo-

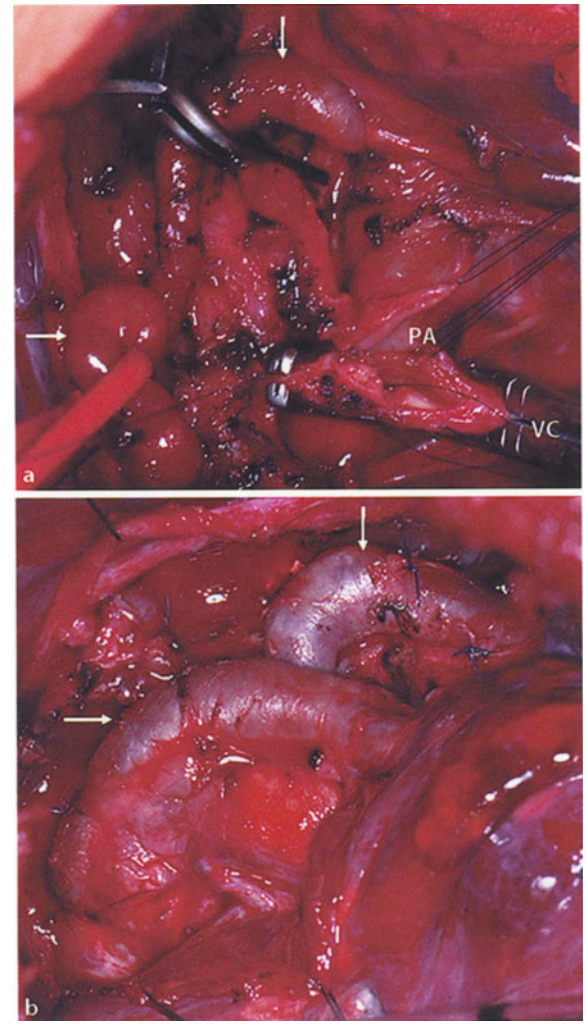


Fig. 2.4.13. Pulmonary atresia with ventricular septal defect: surgery. **a** Preparation of two major aortopulmonary collateral arteries (white arrows) for unifocalization with end-to-side anastomosis to the pulmonary artery branch, longitudinally incised and controlled with a vascular clamp (*PA* pulmonary artery, *VC* vascular clamp), **b** completed anastomosis of the two major aortopulmonary collateral arteries (white arrows) to the pulmonary artery branch

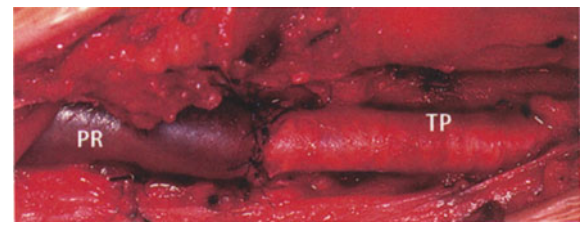
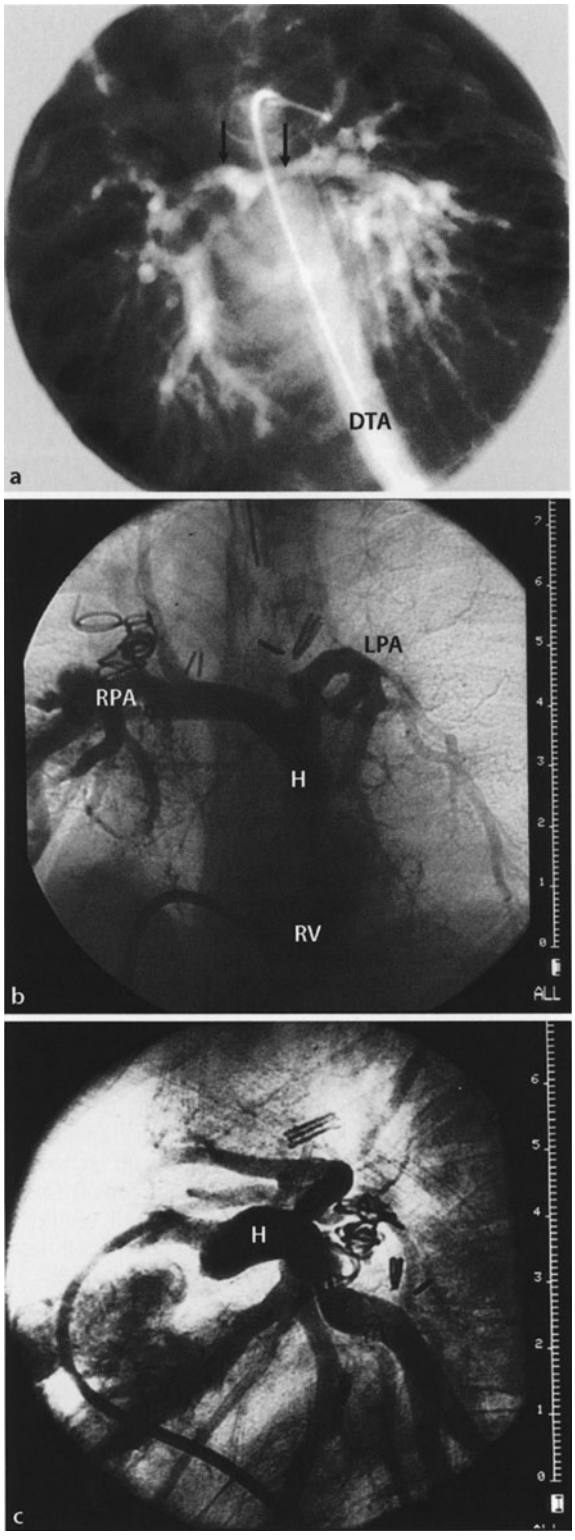


Fig. 2.4.14. Pulmonary atresia with ventricular septal defect: surgery. Intraoperative photograph after unifocalization with a pericardial roll and central shunt with a PTFE tubular prosthesis (*PR* pericardial roll, *TP* tubular prosthesis)



nary artery reconstructed confluence. The ventricular septal defect is left open, or it may be closed with a fenestrated PTFE patch, to allow right ventricular decompression, with later closure using a procedure of interventional cardiology.

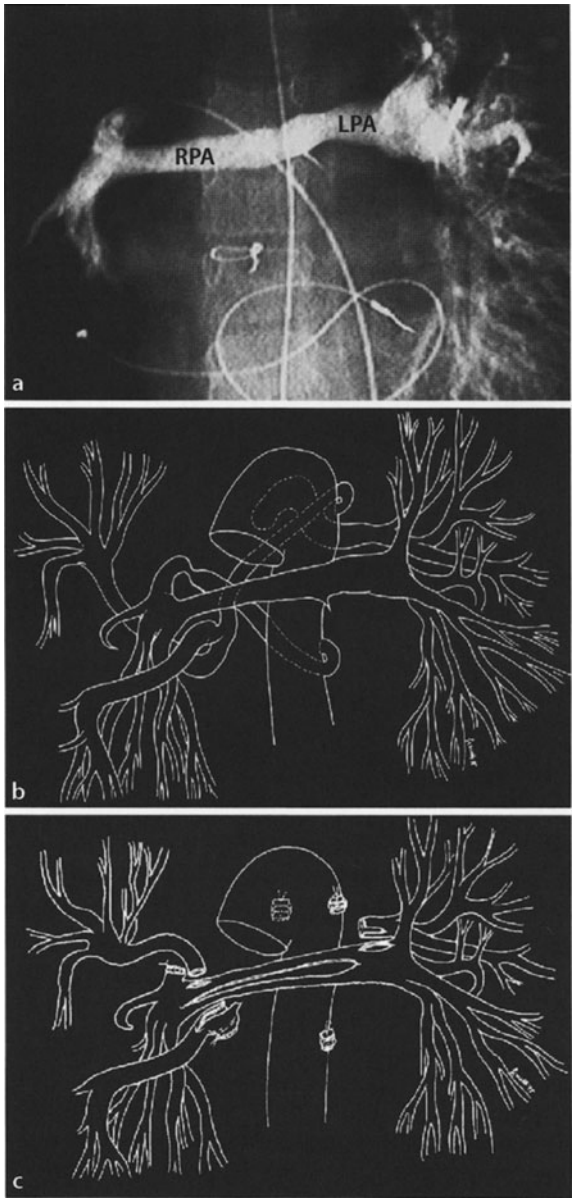
Single-stage unifocalization and complete repair

The unifocalization of the major aortopulmonary collateral arteries is performed as for the two-stages repair (see above); the ventricular septal defect is closed from a longitudinal right ventriculotomy with a prosthetic patch, leaving the aortic valve in communication with the left ventricle; the right ventricle to pulmonary artery continuity is obtained with a biological valved conduit (rarely a monocusp valve patch or a valveless conduit is used) interposed between the same right ventriculotomy used for closure of the ventricular septal defect and the pulmonary artery reconstructed confluence.

Alternative surgical approach

The segment of the descending thoracic aorta containing the origin of all the collaterals, reached on cardiopulmonary bypass through a median sternotomy, is transected during a period of circulatory arrest and replaced

Fig. 2.4.15. Pulmonary atresia with ventricular septal defect: surgery. **a** Preoperative angiography in a neonate with injection in the descending thoracic aorta showing multiple aortopulmonary collateral arteries retrogradedly perfusing the true, confluent, hypoplastic pulmonary arteries (white arrows), with the typical imaging of "sea gull" at their confluence, **b** angiography in the anteroposterior view of the same patient at 8 years of age, after unifocalization and implantation of an homograft between the right ventricle and the pulmonary arteries bifurcation, leaving the ventricular septal defect open; later this patient underwent final repair, with closure of the ventricular septal defect (*H* homograft, *LPA* left pulmonary artery, *RPA* right pulmonary artery, *RV* right ventricle, **c** angiography in the lateral view showing opacification of the pulmonary artery branching for both lungs



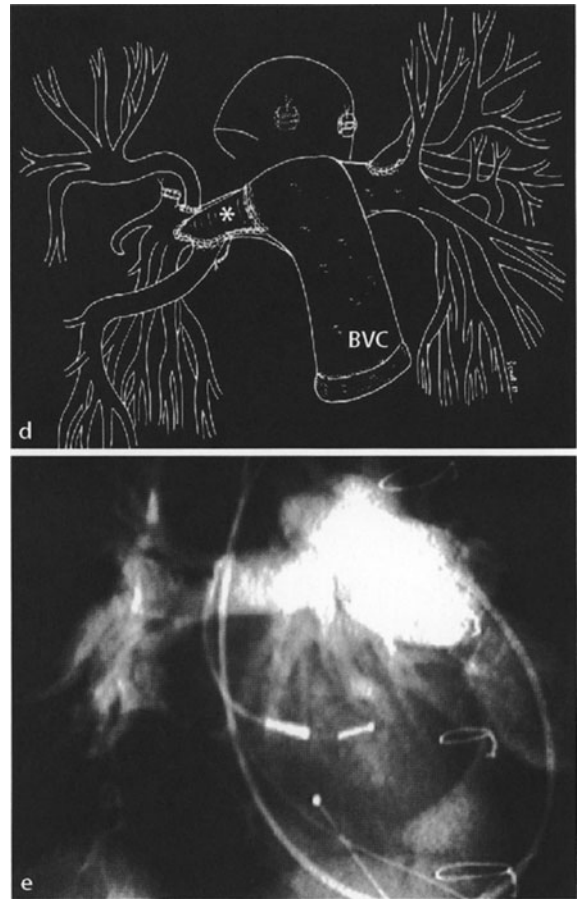
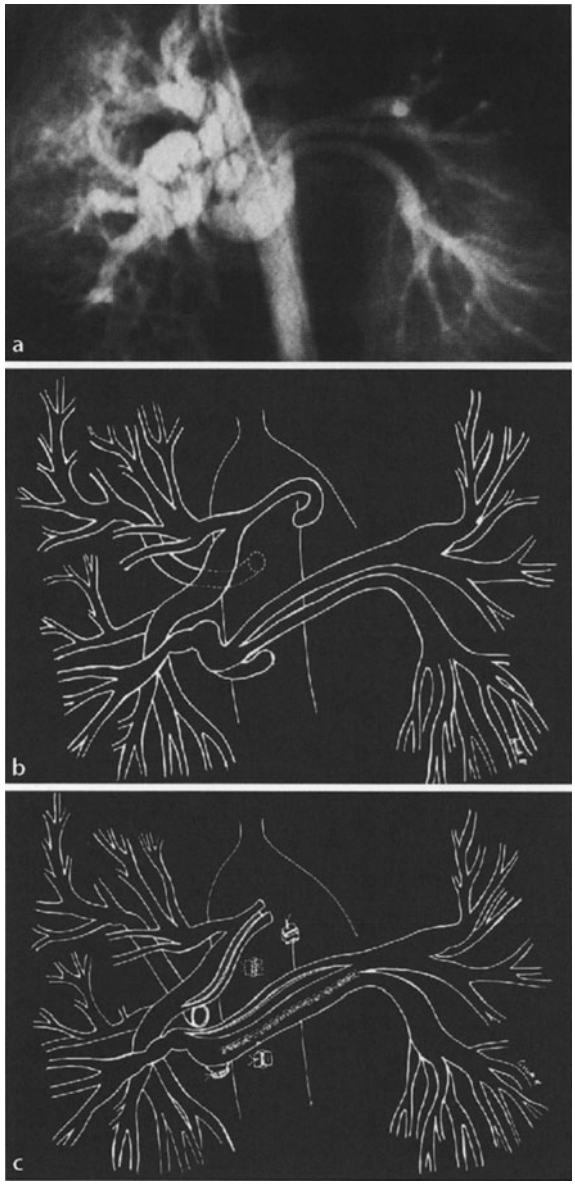
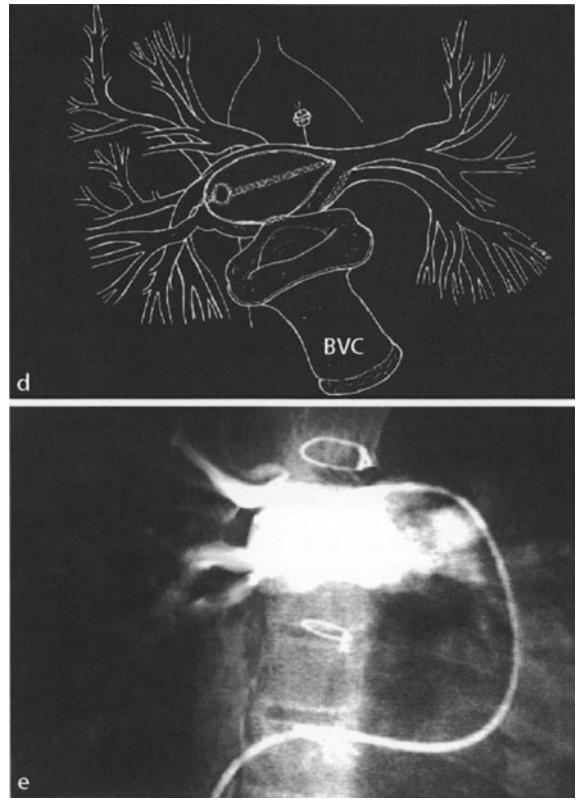


Fig. 2.4.16. Pulmonary atresia with ventricular septal defect: surgery. **a** Preoperative angiography in the anteroposterior view in a child with pulmonary atresia, ventricular septal defect and major aortopulmonary collateral arteries, showing the presence of native pulmonary arteries of intermediate size (*LPA* left pulmonary artery, *RPA* right pulmonary artery) (photograph courtesy of Dr. Adriano Carotti), **b** schematic drawing of the native pulmonary arteries and major aortopulmonary collaterals corresponding to the angiography in **a**, **c** schematic drawing of the surgical unifocalization: the major aortopulmonary collaterals are closed with double pledget-supported sutures at their aortic origin, and then transected and longitudinally opened; the native pulmonary arteries are longitudinally opened, **d** schematic drawing of the final result of the surgical unifocalization, with posterior tissue-to-tissue anastomosis and anterior augmentation with autologous pericardial patch (white star); the continuity between the right ventricle and the pulmonary artery confluence is obtained with a biological valved conduit (*BVC* biological valved conduit), **e** postoperative angiography in the same child showing the final result of the surgical unifocalization, with totally unifocalized pulmonary arterial tree (**b–e** reproduced with permission from: Carotti A, di Donato RM, Squitieri C, Guccione P, Catena G (1998) Total repair of pulmonary atresia with ventricular septal defect and major aortopulmonary collaterals: an integrated approach. J Thorac Cardiovasc Surg 116:914–923)





d schematic drawing of the final result of the surgical unifocalization, with posterior tissue-to-tissue anastomosis and anterior augmentation with the distal and of a biological valved conduit (*BVC* biological valved conduit), **e** postoperative angiography in the same child showing the final result of the surgical unifocalization, with a totally unifocalized pulmonary aterial tree (**b–e** reproduced with permission from Carotti A, di Donato RM, Squitieri C, Guccione P, Catena G (1998) Total repair of pulmonary atresia with ventricular septal defect and major aortopulmonary collaterals: an integrated approach. J Thorac Cardiovasc Surg 116:914–923)

Fig. 2.4.17. Pulmonary atresia with ventricular septal defect: surgery. **a** Preoperative angiography in the anteroposterior view in a child with pulmonary atresia, ventricular septal defect and major aortopulmonary collateral arteries, showing the absence of native pulmonary arteries (photograph courtesy of Dr. Adriano Carotti), **b** schematic drawing of the major aortopulmonary collaterals corresponding to the angiography in **a**, **c** schematic drawing of the surgical unifocalization: the major aortapulmonary collaterals are closed with double pledget-supported sutures at their aortic origin, and then transected and longitudinally opened,

with a PTFE conduit. The distal end of the above segment is closed by running suture, while the proximal end is end-to-side anastomosed to the native pulmonary artery confluence. At this point the conventional intracardiac repair is performed, with patch closure of the ventricular septal defect and interposition of a biological valved conduit between the right ventricle and the confluence between the native pulmonary arteries and the transected aortic segment.

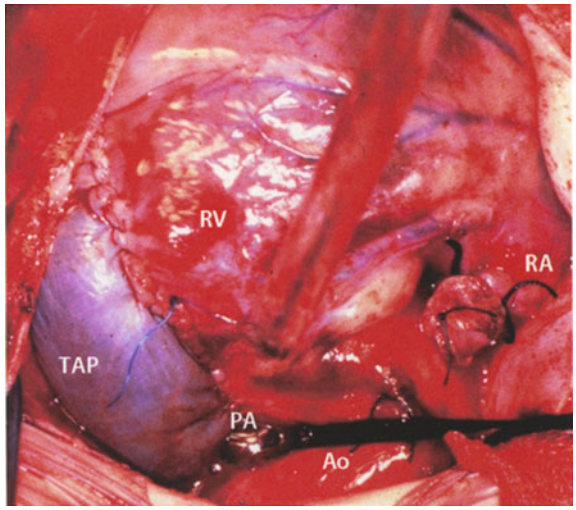


Fig. 2.4.18. Pulmonary atresia with ventricular septal defect: surgery. Intraoperative photograph showing the end of the surgical procedure consisting of closure of the ventricular septal defect and transannular heterologous pericardial patch. The ascending aorta is displaced on the right side to show the distal end of the patch (*Ao* aorta, *PA* pulmonary artery, *RA* right atrium, *RV* right ventricle, *TAP* transannular patch)

Potential complications

Inadequate and/or dishomogeneous distribution of pulmonary blood flow; congestive heart failure, due to the persistence of uncontrolled major aortapulmonary collateral arteries; residual or recurrent ventricular septal defect; complete atrioventricular block, ventricular or supraventricular arrhythmias; persistent airway hyperresponsiveness with or without bronchomalacia; bronchospasm due to tracheobronchial epithelial necrosis or ischemia due to the airway ischemia resulting from interruption of the tracheobronchial blood supply during dissection and mobilization of major aortopulmonary collaterals; development of pulmonary vascular obstructive disease; development or progressive increase of aortic valve regurgitation; need for biological conduit replacement because of conduit calcification and/or valve degeneration (=pulmonary valve regurgitation) or because patient outgrew the conduit size (=right ventricular outflow tract obstruction).

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Chapter 2.5 Truncus arteriosus

Incidence

Truncus arteriosus is a rare congenital heart defect, with a reported incidence between 0.16 and 1.27 every 10,000 newborns, or 1.4–2.8% of congenital heart defects.

Morphology (Fig. 2.5.1)

There is only one great artery or trunk as blood exits from the base of the heart, which then branches giving origin to the coronary arteries, the pulmonary arteries and the systemic circulation (Fig. 2.5.2).

This great artery (truncus arteriosus) usually has one semilunar valve (truncal valve) which may have between one and six leaflets: a truncal valve with one, five or six leaflets is extremely rare, while the most frequent type is a truncal valve with three leaflets (42–61% of cases), followed by the truncal valve with four leaflets (24–31% of cases) and with two leaflets (5% of cases). The semilunar truncal valve is generally located above the ventricular septal defect, almost always present, and can present with dysplastic thickened and deformed leaflets, with resulting stenosis, regurgitation (in at least 20% of cases), or both (Fig. 2.5.3).

The ventricular septal defect results from the absence of the infundibular septum, and is generally high, anterior and unrestrictive. Absence of the ventricular septal defect is anedoctical.

The pulmonary arteries can originate as a single main pulmonary artery from the lateral aspect of the truncus arteriosus (type I

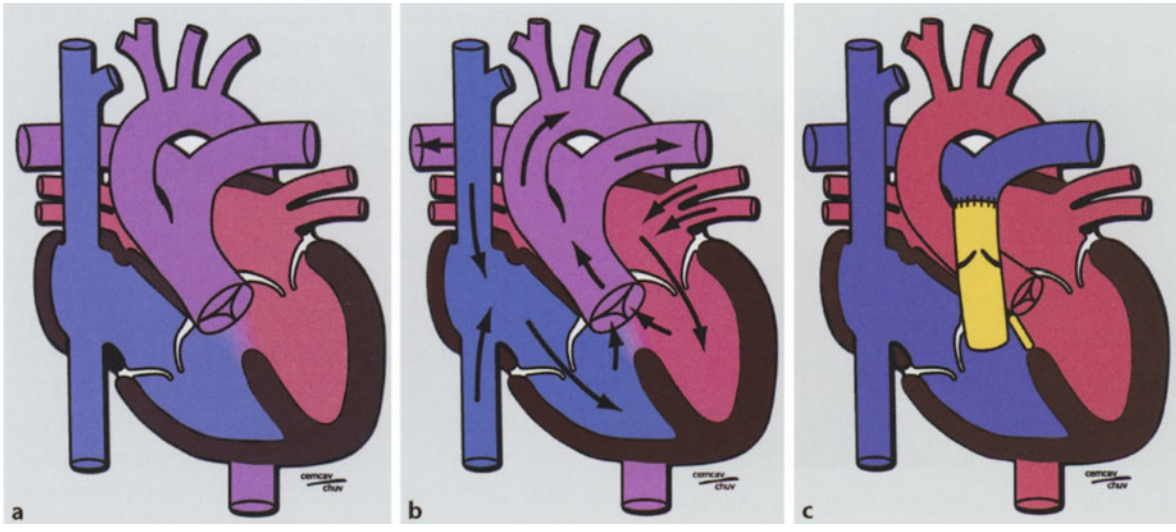

Fig. 2.5.1. Truncus arteriosus. **a** Morphology, **b** pathophysiology, **c** surgery

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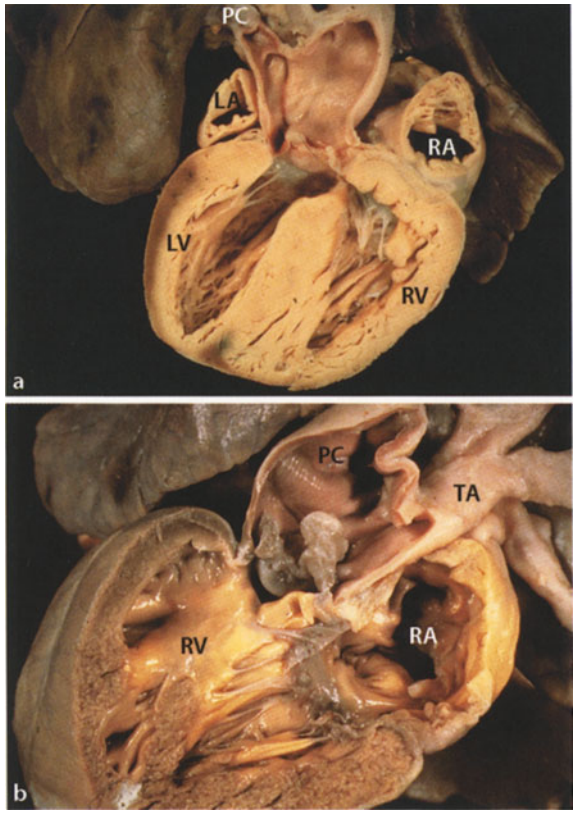


Fig. 2.5.2. Truncus arteriosus: morphology. **a** Posterior view of the truncus arteriosus, with the pulmonary component originating from the left lateral side of the truncus (*LA* left atrium, *LV* left ventricle, *PC* pulmonary component, *RA* right atrium, *RV* right ventricle) (photograph courtesy of Dr. Bruno Marino), **b** anterolateral aspect of the truncus arteriosus, with the pulmonary component originating from the left lateral side of the truncus (reproduced with permission from: Marino B, Thiene G (1990) Atlante di anatomia ecocardiografica delle cardiopatie congenite, USES, Firenze)

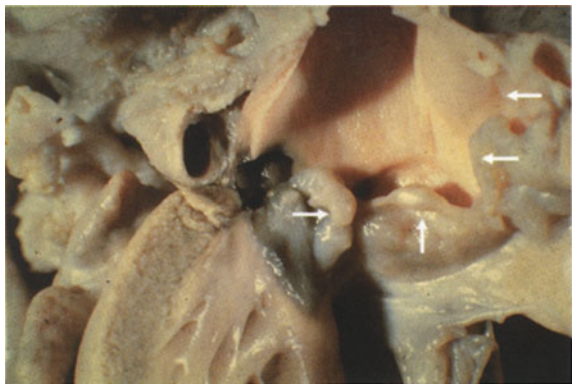


Fig. 2.5.3. Truncus arteriosus: morphology. Aspect of the truncal valve, with four leaflets (white arrows) (photograph courtesy of Dr. Bruno Marino)

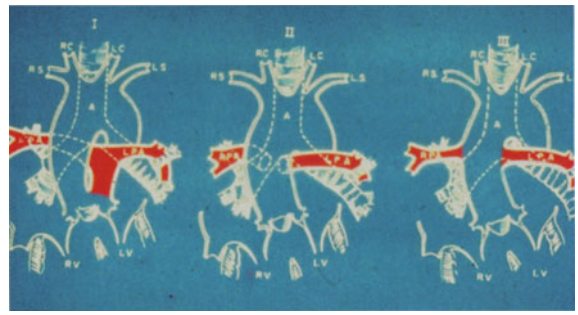


Fig. 2.5.4. Truncus arteriosus: morphology. Collett and Edwards classification in types I, II and III (*A* aorta, *LC* left carotid artery, *LPA* left pulmonary artery, *LS* left subclavian artery, *LV* left ventricle, *RC* right carotid artery, *RPA* right pulmonary artery, *RS* right subclavian artery, *RV* right ventricle (photograph courtesy of Dr. Carlo Marcelletti)

of the Collett and Edwards classification, the most frequent), or with two very close but separate origins from the posterolateral aspect of the truncus arteriosus (type II, the second frequent), or with the two pulmonary arteries independently originating from a lateral aspect of the truncus arteriosus (type III) (Fig. 2.5.4). The truncus arteriosus type IV, with neither pulmonary arterial branch arising from the truncus arteriosus, is not considered as a true entity, but only as a variant of pulmonary atresia with ventricular septal defect and absent pulmonary arteries (see chapter "Pulmonary atresia with ventricular septal defect"). Stenosis at the origin of one or both branches of the pulmonary arteries is reported in 2 to 10% of patients.

The origin of coronary arteries presents with a variable pattern, independent of the number of truncal valve leaflets.

Associated anomalies

Ventricular septal defect is almost always present, right aortic arch is frequent (18–36%), as well as truncal valve regurgitation (23%) or stenosis and coronary arteries abnormalities (18%), including single coronary artery and intramural course; aortic arch interruption (Fig. 2.5.5), usually of type B (11–14%), and aortic coarctation are accompanied by

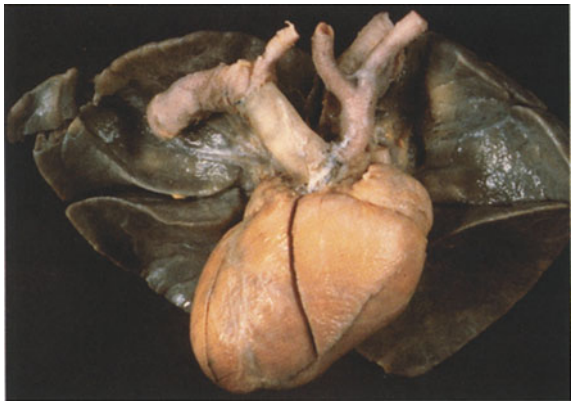


Fig. 2.5.5. Truncus arteriosus: morphology. Truncus arteriosus with aortic arch interruption (posterior aspect) (photograph courtesy of Dr. Bruno Marino)

patent ductus arteriosus; more rare (5%) is the association with non-confluent pulmonary arteries, the so-called "absent" pulmonary artery, where one of the pulmonary arteries in reality originates either from a patent ductus arteriosus or from a major aortopulmonary collateral; persistent left superior vena cava, total anomalous pulmonary venous connection, tricuspid atresia, complete atrioventricular septal defect, single ventricle, double aortic arch, anomalous origin of circumflex coronary artery from the right pulmonary artery, situs inversus, dextrocardia have been exceptionally reported.

Approximately 35% of children with truncus arteriosus have microdeletions within chromosome band 22q11.2, while 30–35% present with DiGeorge (=velocardiofacial syndrome with deletion of chromosome 22q11.2, previously called CATCH-22=cardiac defect, abnormal face, thymic hypoplasia, cleft palate, hypocalcemia, microdeletion of band 22q11) syndrome.

Pathophysiology

With the exception of the rare group (less than 10%) with obstruction to the pulmonary blood flow due to stenosis of the origin of the pulmonary artery component, generally the decrease of pulmonary vascular resistance typical of the first few weeks of life is accompanied by an increase in the total pulmonary blood flow, frequently up to extreme levels, with low diastolic and mean aortic pressure and the consequent reduced coronary artery perfusion pressure. The clinical consequences of this pathophysiologic pattern are cyanosis (due to the mixing of desaturated and oxygenated blood in the truncus arteriosus), congestive heart failure (due to the huge left ventricular volume overload and right ventricular pressure overload) and myocardial ischemia (due to the coronary arteries with the left-to-right shunt towards the pulmonary circulation). The result of pulmonary overcirculation and increased myocardial work is an increased resting oxygen demand and decreased metabolic reserve, with progressive metabolic acidosis and myocardial dysfunction.

Diagnosis

Clinical pattern: symptoms are variable, depending upon the specific anatomic features and age at presentation:

- poor feeding, tachypnea, tachycardia, shortness of breath, wheezing, grunting, nasal flaring, restlessness, liver distension, neck vein distension,
- facial swelling, mild to moderate cyanosis, bounding peripheral pulses;
- respiratory distress can be present, usually due to the airway compression;
- cardiac signs include hyperdynamic precordium, aortic ejection click, single second sound (due to the single outlet); respectively conspicuous diastolic murmur or loud systolic ejection murmur characterize the presence of severe regurgitation or stenosis of the truncal valve.

Electrocardiogram: right, left or biventricular hypertrophy, with or without ST-T changes, depending on the presence of truncal valve abnormalities and/or myocardial ischemia; left atrial enlargement present with substantial pulmonary overcirculation.

Chest X-ray: cardiomegaly, absence of the pulmonary trunk segment, increased pulmonary arterial vascularity, lung hyperinflation; right aortic arch (18–36% of cases) can be accompanied by airway compression.

Echocardiogram: diagnostic in almost all cases, it provides all the information needed for the surgical decision (Fig. 2.5.6); the subcostal and parasternal long-axis views show the single great artery arising from the ventricles, with variable override of the ventricular septum, and origin(s) of the pulmonary arteries from the truncus arteriosus; the parasternal short-axis view shows the morphology and function (with Doppler color flow) of the truncal valve and origin and course of the proximal coronary arteries; the high parasternal and suprasternal views define the position of the aortic arch.

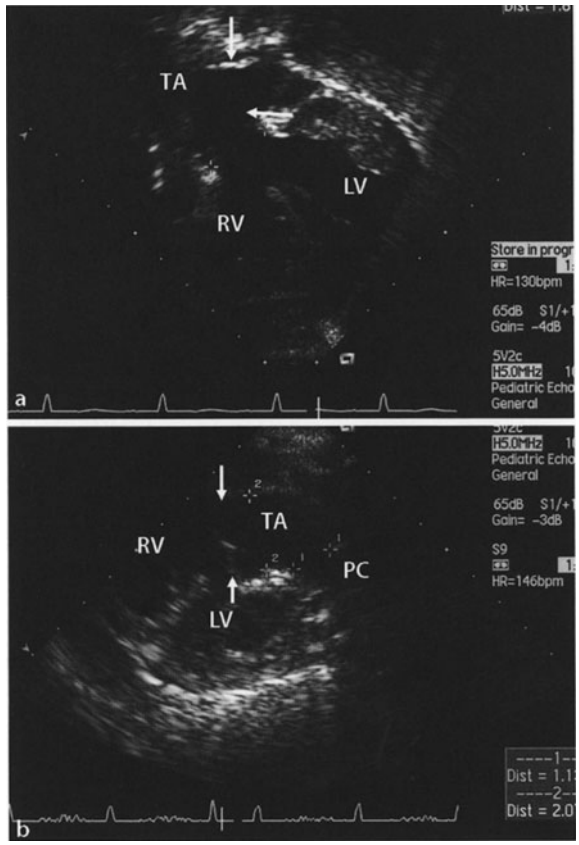


Fig. 2.5.6. Truncus arteriosus: echocardiography. **a** Subxiphoid view showing the origin of the truncus arteriosus above a very large ventricular septal defect, and the origin of the pulmonary artery component (white arrows), **b** long-axis view in the same patient showing the origin of the truncus arteriosus with the truncal valve (white arrows), and the origin of the pulmonary artery component (*LV* left ventricle, *RV* right ventricle, *PC* pulmonary artery component, *TA* truncus arteriosus) (photographs courtesy of Dr. Stefano di Bernardo)

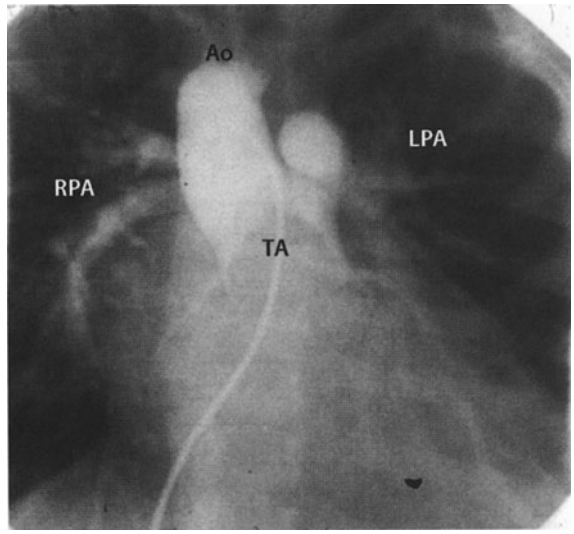


Fig. 2.5.7. Truncus arteriosus: angiography. Anteroposterior view of a contrast injection in the root of the truncus arteriosus, giving origin to the three components: aorta, pulmonary artery and coronary arteries (*Ao* aorta, *LPA* left pulmonary artery, *RPA* right pulmonary artery, *TA* truncus arteriosus)

Cardiac catheterization: poorly tolerated in very sick neonates, is performed only in the case of unusual anatomy on the echocardiography and of unusually associated defects (Figs. 2.5.7 and 2.5.8); in older infants to quantify the degree of pulmonary vascular resistance or to rule out pulmonary vascular obstructive disease in older children.

Indications for surgical treatment

This congenital heart defect, untreated, carries a very high mortality, because of congestive heart failure and myocardial ischemia in early infancy; mortality without treatment is 50% in the first month of life, 80% in the first three months. Therefore indication for surgery is generally given within

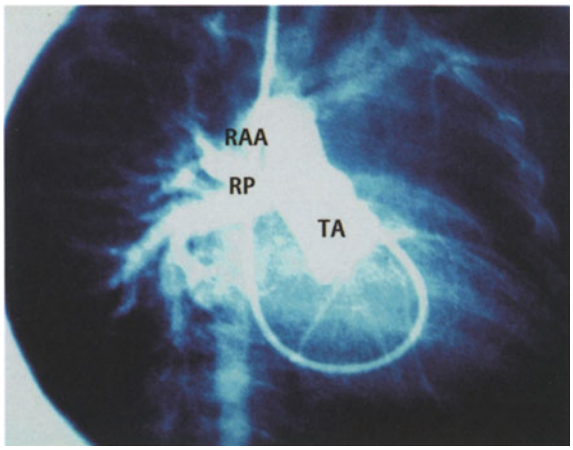


Fig. 2.5.8. Truncus arteriosus: angiography. Anteroposterior view of a contrast injection in the root of the truncus arteriosus, giving origin to the aorta with right aortic arch, right pulmonary artery and coronary arteries; the left pulmonary artery is not visualized (*RAA* right aortic arch, *RPA* right pulmonary artery, *TA* truncus arteriosus)

the first few weeks of life, once the general condition of the neonate have been stabilized with the medical control of congestive heart failure and with hyperalimentation either intravenous or via a nasogastric feeding. In older infants and children the indication for surgery is given provided that the presence of pulmonary vascular obstructive disease has been ruled out.

In borderline cases with elevated pulmonary vascular resistance the response to intravenous or inhaled pulmonary vasodilators can be measured during cardiac catheterization. Extremely rare is the presentation of an older patient who survived with the development of pulmonary vascular obstructive disease. Challenging is the decision how to manage an associated lesion of the truncal valve (very frequent), because in the presence of truncal valve stenosis as well as of truncal valve regurgitation of moderate degree it is quite difficult to predict the hemodynamic pattern obtained at the level of the truncal valve after repair, when the flow across the truncal valve itself (which becomes aortic valve) will be limited to the systemic output and not to the preoperative combination of elevated pulmonary and systemic blood flow.

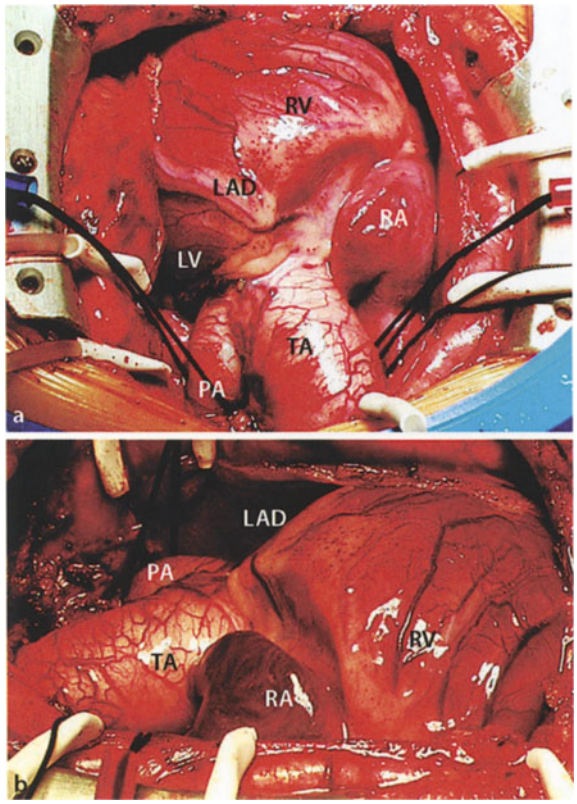
Surgical treatment (on cardiopulmonary bypass)

The palliative approaches with bilateral pulmonary artery banding or plication of the origin of the pulmonary artery component have been practically abandoned in favor of the surgical repair.

The surgical technique (Fig. 2.5.9) consists of three major components:

- the separation of the pulmonary artery component from the truncus; at this point the truncus becomes the new aorta; the remaining opening in the lateral aspect of the truncus can be closed either directly or with a patch (pericardium or PTFE), accordingly with the size and particularly with the presence of the origin of a coronary artery in proximity of the opening itself; either technique of closure of the residual opening (direct or patch) needs to avoid traction or tension on the coronary arteries, and in this regard patch closure is the preferable technique;
- the closure of the ventricular defect with a patch from a longitudinal right ventriculotomy, leaving the remaining systemic artery (becoming the aorta) in connection with only the left ventricle, and avoiding damage to the truncal valve (becoming aortic valve);
- the connection of the pulmonary artery component with the right ventricle by interposition of a biological valved conduit between the distal pulmonary artery component and the right ventriculotomy; valveless conduits are more rarely used as the connection between the right ventricle and pulmonary artery; alternatively it is possible to perform a direct anastomosis of the posterior wall of the pulmonary component to the posterior edge of the right ventriculotomy, completing the connection with a pericardial or PTFE roof, with or without a monocusp pulmonary valve (technique rarely utilized).

Associated lesions need to be treated during the same procedure, particularly the



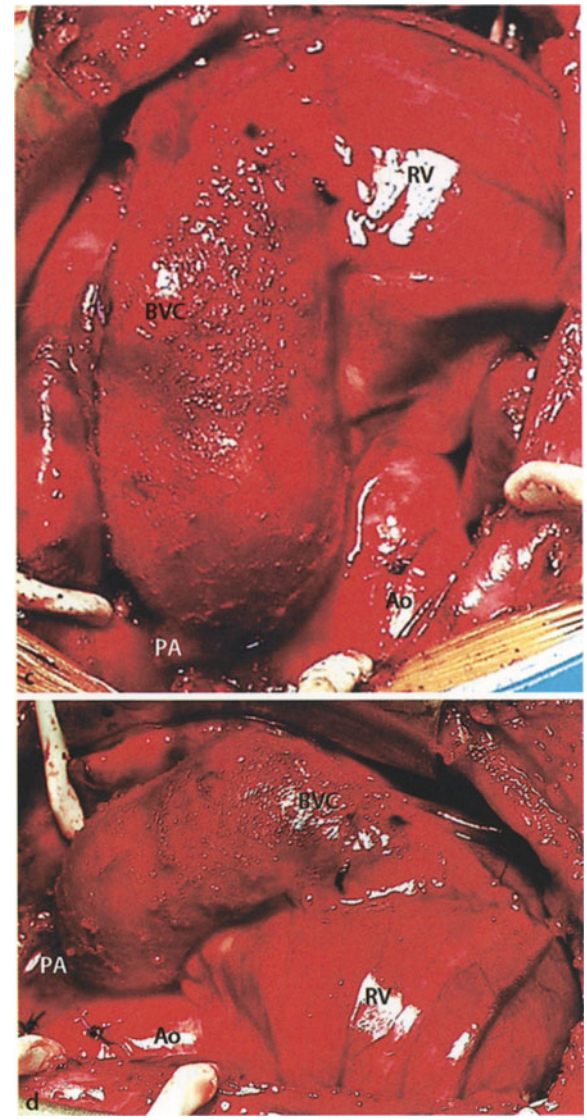


Fig. 2.5.9. Truncus arteriosus: surgery. Intraoperative photograph in a neonate **a** showing the preparation for repair, with both branches of the pulmonary component already controlled by tourniquets (*LAD* left anterior descending coronary artery, *LV* left ventricle, *PA* pulmonary artery, *RA* right atrium, *RV* right ventricle, *TA* truncus arteriosus), **b** taken from the right side, **c** after repair with a 16 mm Contegra biological valved conduit interposed between the right ventricle and pulmonary component (*Ao* aorta, *BVC* biological valved conduit), **d** after repair taken from the right side

presence of aortic coarctation or aortic arch interruption.

In the presence of moderate to severe truncal valve regurgitation, particularly with a quadrileaflet valve, a plasty of reduction to three leaflets with leaflet excision and annular remodeling can substantially reduce the degree of valvular regurgitation. More difficult to perform is the surgical opening of a stenotic truncal valve with adequate results. Truncal valve replacement (with a homograft or mechanical valve) needs to be taken into consideration in case of failure of the reconstructive valvular surgery.

Potential complications

Residual ventricular septal defect, complete atrioventricular block, arrhythmias, residual or progressive truncal valve stenosis or regurgitation, residual right ventricular outflow tract obstruction, pulmonary hypertension, airways compression (particularly in the presence of right aortic arch and aortic coarctation or aortic arch interruption). Late complications are progressive aortic (previous truncal) valve dysfunction and obstruction of the right ventricle to pulmonary artery conduit, due in most cases to calcification and/or patient overgrowth.

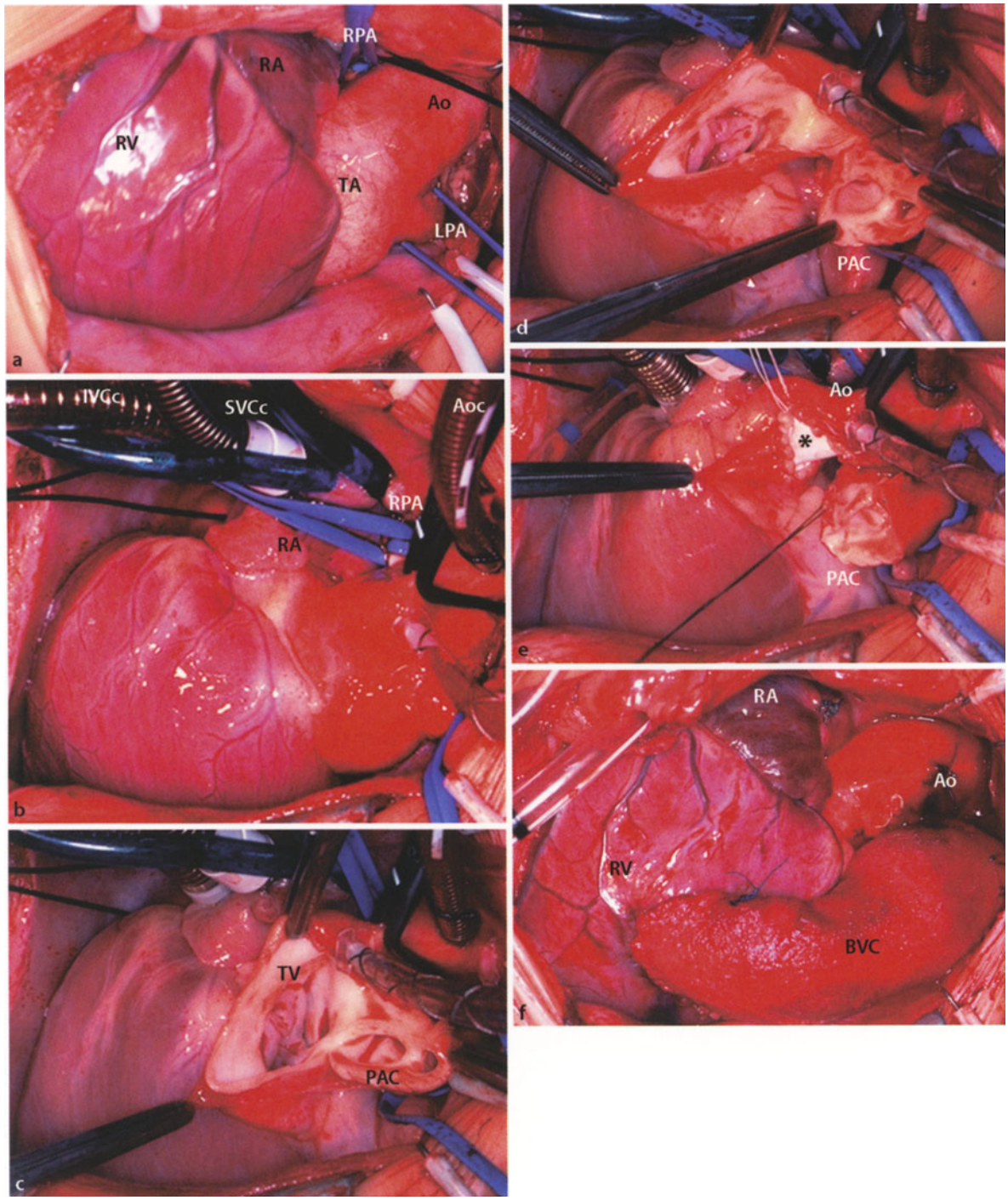


Fig. 2.5.10. Truncus arteriosus: surgery. **a** Intraoperative photograph in the child of Fig. 2.5.6 showing the anatomy of the truncus arteriosus, with the two pulmonary arteries already controlled with blue elastic vessel loops (*Ao* aorta, *LPA* left pulmonary artery, *RA* right atrium, *RPA* right pulmonary artery, *RV* right ventricle, *TA* truncus arteriosus), **b** Truncus arteriosus prepared for repair, with establishment of cardiopulmonary bypass, snaring of the pulmonary arteries, aortic cross clamping and cardioplegia administration (*Aoc* aortic cannula, *CPc* cardioplegia cannula, *IVCc* inferior vena cava cannula, *SVCc* superior vena cava cannula), **c** truncus arteriosus opened, with initial separation of the origin of the pulmonary artery component, and visualization of the truncal valve and of the origin of the left coronary artery immediately underneath the pulmonary artery component (*PAC* pulmonary artery component, *TV* truncal valve), **d** completed separation of the origin of the pulmonary artery component, **e** closure of the residual opening in the truncus arteriosus (now aorta) after separation of the pulmonary component with PTFE patch (black asterisk), **f** photograph after final repair with a 18 mm Contegra biological valved conduit interposed between the right ventricle and pulmonary component (*Ao* aorta, *BVC* biological valved conduit)

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Chapter 2.6 Aortopulmonary window

Incidence

Aortopulmonary window is a rare congenital malformation, with an incidence of 0.2% in children with congenital heart defects.

Morphology (Fig. 2.6.1)

Aortopulmonary window is a communication, usually nonrestrictive, with a more or less circular border, between the ascending aorta and the main pulmonary artery, in the presence of two semilunar valves. In most patients there is little or no length to the communication, as the term window implies, while a ductus-like type of communication is rare.

The aortopulmonary window is generally located in the left lateral wall of the ascending aorta, usually close to the orifice of the left coronary artery; therefore it is not infrequent to find an anomalous origin of the coronary artery from the pulmonary artery, close to the edge of the defect.

Associated anomalies

Associated defects are present in 30–50% of patients, the most frequent being aortic arch interruption type A, tetralogy of Fallot with or without pulmonary atresia, ventricular septal defect, anomalous origin of a coronary artery and anomalous origin of a pulmonary artery; occasionally an association has been reported with atrial septal defect, cor triatriatum, complete atrioventricular septal defect, transposition of the great ar-

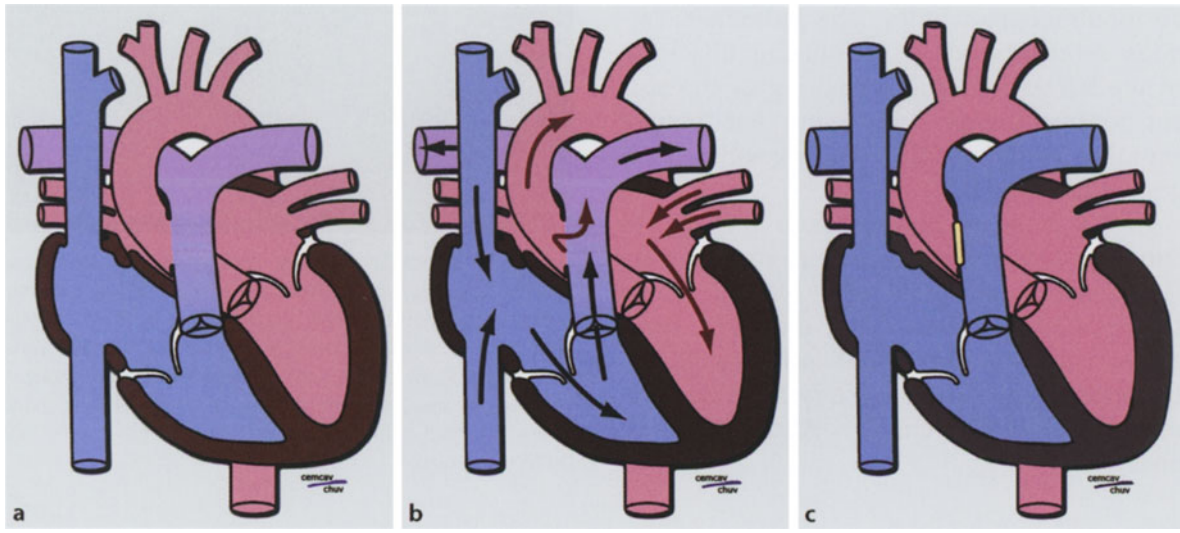


Fig. 2.6.1. Aortopulmonary window. **a** Morphology, **b** pathophysiology, **c** surgery

A. F. Corno, *Congenital Heart Defects*

teries, subaortic obstruction (extremely rare), bicuspid aortic valve, aortic atresia, critical pulmonary valve stenosis, aortic arch hypoplasia, aortic coarctation, aortic arch interruption, double aortic arch, and patent ductus arteriosus.

Pathophysiology

The pathophysiology is in large part determined by the size of the defect and by the presence and type of associated lesions. In the presence of defect of small size (about 10% of the patients) the pathophysiology is very similar to the one observed in patients with patent ductus arteriosus (see chapter "Patent ductus arteriosus"). If the defect is large, as in the vast majority of patients, the consequence is a large left-to-right shunt with pulmonary hypertension, and early development of pulmonary vascular obstructive disease. This pattern is even increased in the presence of the frequently associated obstructive anomalies of the aortic arch.

Like all arterial-level shunts, flow occurs during both systole and diastole; the magnitude depending on both the size of the aortopulmonary window and the relative resistance of pulmonary and systemic circulations. In the first few months of life, due to the left-to-right shunting through a large aortopulmonary window, the pulmonary venous return is increased causing dilatation of the left atrium and stretching of the patent foramen ovale with more left-to-right shunting at the atrial level, worsening congestive heart failure.

In the presence of anomalous origin of a coronary artery, the anomalous coronary artery remains perfused with systemic pressure and oxygen saturation; therefore, there are no signs of myocardial ischemia like in the anomalous coronary artery from the pulmonary artery (see chapter "Anomalous coronary arteries").

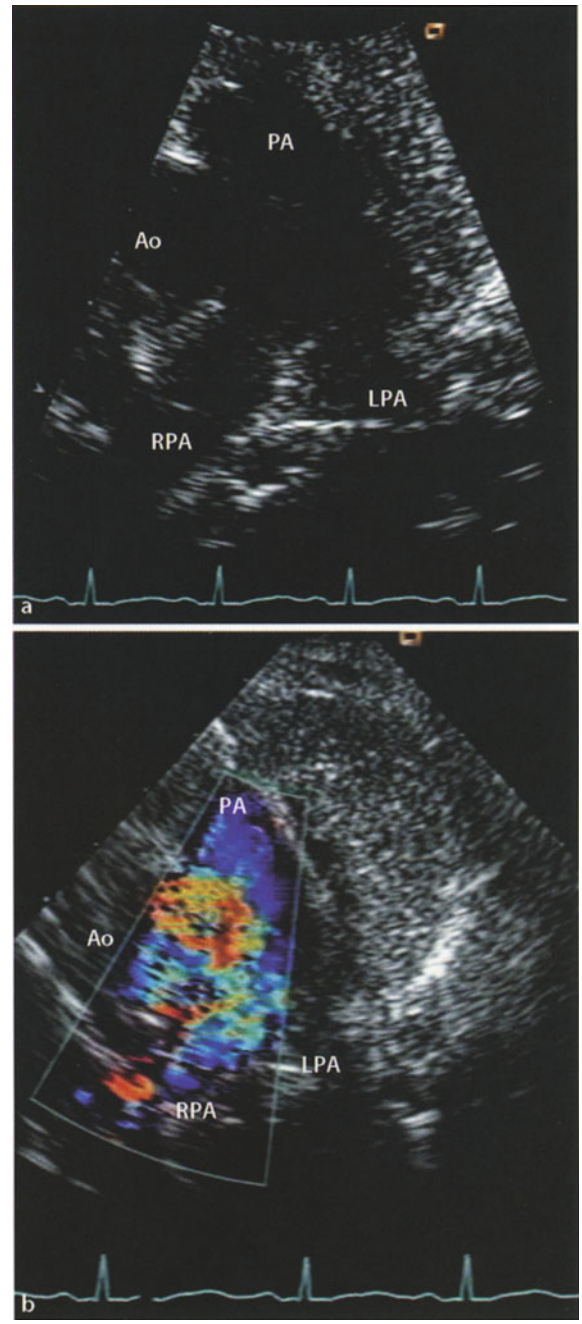


Fig. 2.6.2. Echocardiography. **a** Parasternal short-axis view showing the large aortopulmonary window (white arrows), **b** parasternal short-axis view showing view of the same patient as in **a** showing the color Doppler investigation with left-to-right shunt through the unrestrictive aortopulmonary window (*Ao* aorta, *LPA* left pulmonary artery, *PA* pulmonary artery, *RPA* right pulmonary artery) (photographs courtesy of Dr. Nicole Sekarski)

Diagnosis

Clinical pattern: *small defect*: asymptomatic, or recurrent respiratory infections; the cardiac murmur is generally continuous;

large defect: tachypnea, dyspnea, poor feeding, delayed growth, recurrent respiratory infections, congestive heart failure; peripheral pulses are collapsing; the cardiac murmur is generally only systolic, in the third or fourth intercostal space;

Electrocardiogram: right axis deviation, right ventricular hypertrophy, right or combined atrial enlargement.

Chest X-ray: moderate cardiomegaly, with left atrial enlargement; increased pulmonary vascularity or frank pulmonary edema.

Echocardiogram: the diagnosis is readily made with the combination of subcostal right anterior oblique and suprasternal views or with parasternal short-axis view (Fig. 2.6.2 and 2.6.3).

Cardiac catheterization: needed for associated lesions, particularly to visualize the coronary arteries and rule out anomalous origin (Fig. 2.6.4); useful to calculate the pulmonary vascular resistance, and to rule out the presence of pulmonary vascular obstructive disease in older children.

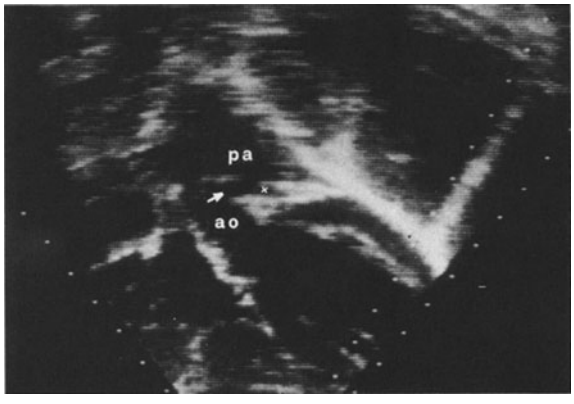


Fig. 2.6.3. Echocardiography. Preoperative echocardiography showing the aortopulmonary window (white arrow) with anomalous origin of the left coronary artery (asterisk) from the ductus-like aortopulmonary window (*Ao* aorta, *PA* pulmonary artery) (reproduced with permission from: Corno AF, Pierli C, Biagioli B, Lisi G, Grossi A (1988) Anomalous origin of the left coronary artery from an aortopulmonary window. J Thorac Cardiovasc Surg 96:669–671)

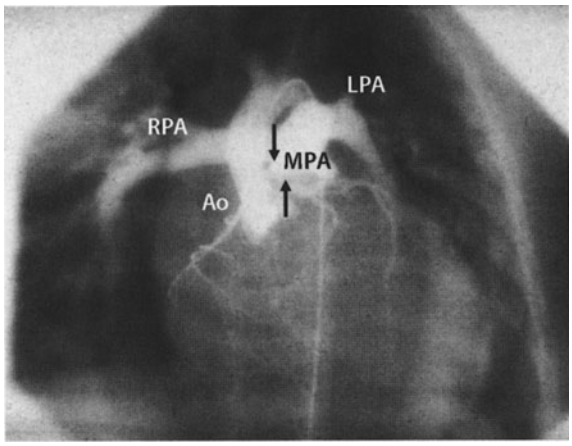


Fig. 2.6.4. Angiography. Anteroposterior view with contrast injection in the ascending aorta showing simultaneous opacification of the pulmonary artery through an aortopulmonary window (black arrows) (*Ao* aorta, *LPA* left pulmonary artery, *MPA* main pulmonary artery, *RPA* right pulmonary artery)

Indications for surgical treatment

While the rare patients with ductus-like aortopulmonary window may remain asymptomatic or oligosymptomatic over a long period, patients with a large aortopulmonary window rarely survive beyond infancy. A large aortopulmonary window could cause pulmonary vascular obstructive disease, if not treated until after two years of age. Therefore indication for surgical closure is generally at the moment of diagnosis, particularly in symptomatic patients.

Surgical treatment (on cardiopulmonary bypass)

Ligature: This procedure, mostly used in the past to obtain closure of the aortopulmonary window without cardiopulmonary bypass, was accompanied by elevated mortality due to the distortion of the origin of a coronary artery originating close to the de-

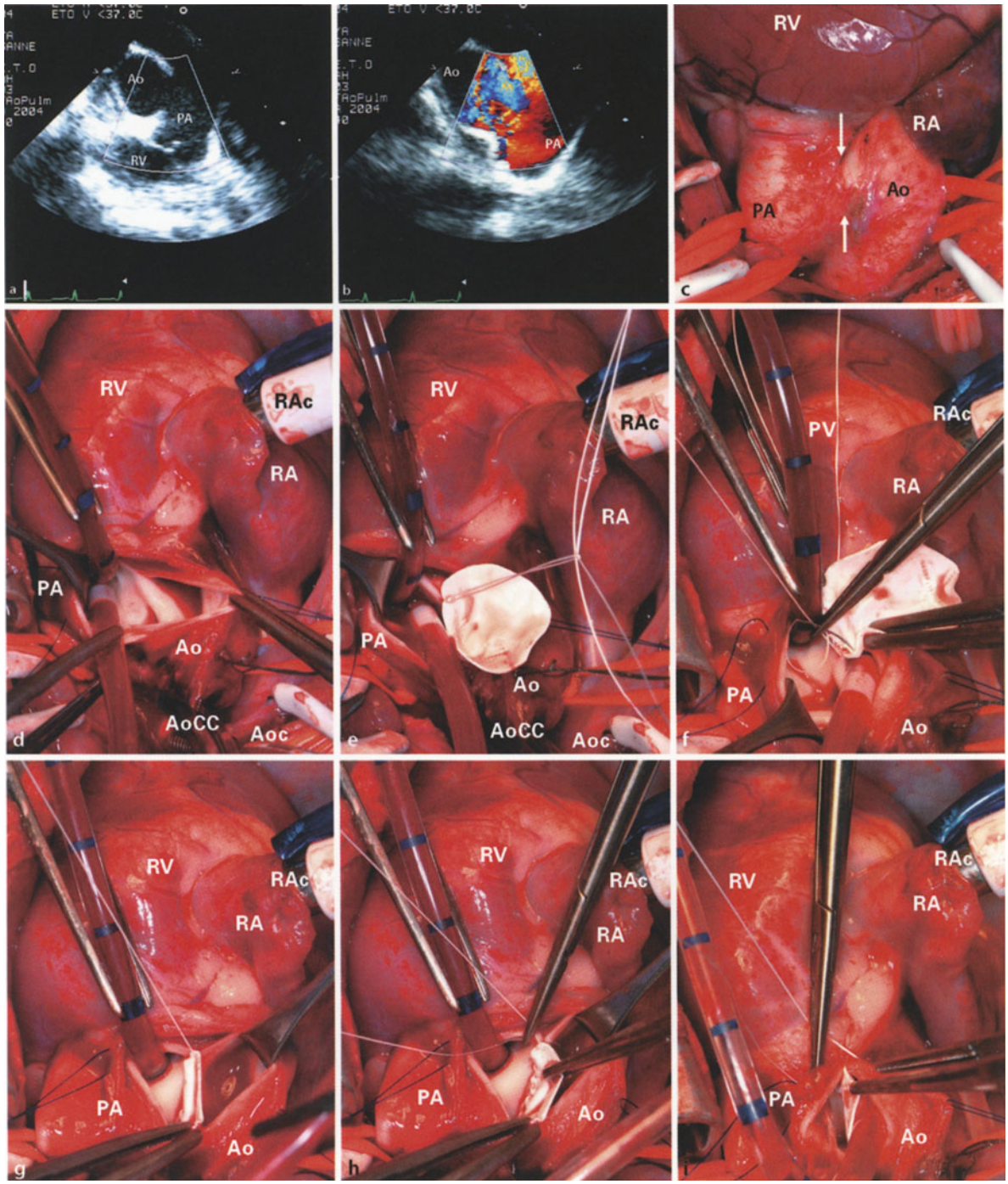
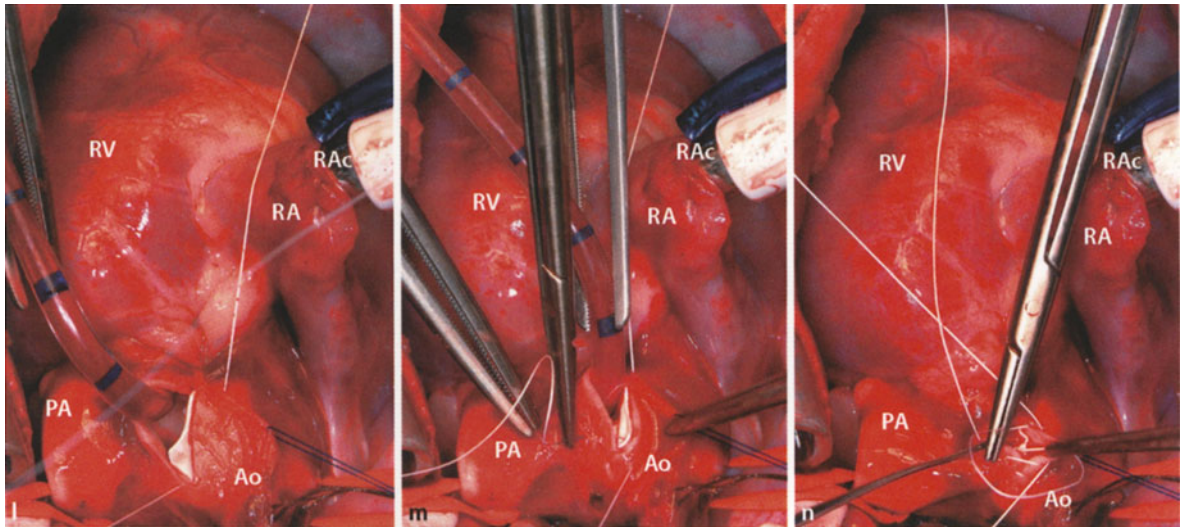
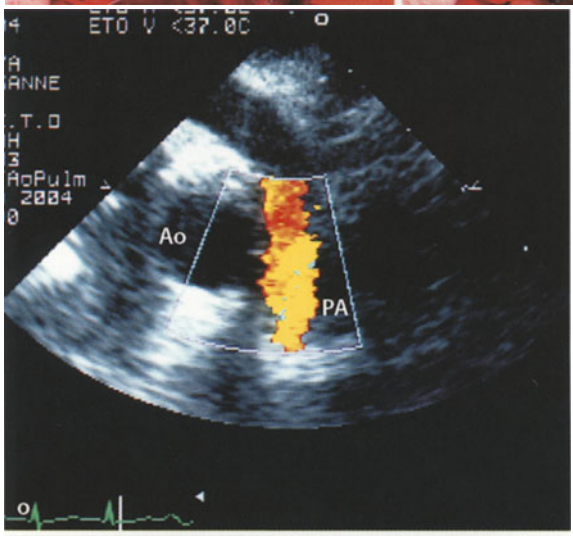
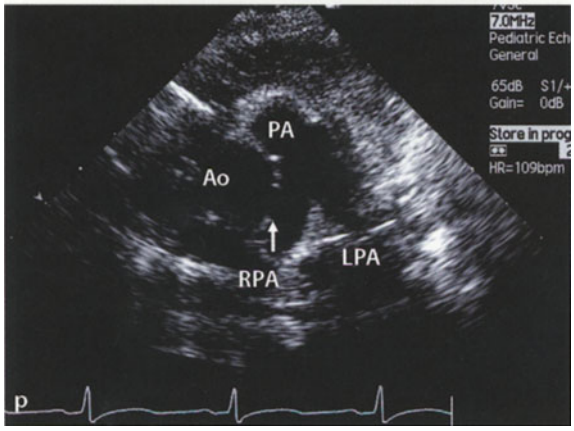


Fig. 2.6.5. Aortopulmonary window: surgery. **a** Intraoperative transesophageal echocardiography, showing the aortopulmonary window of the same patient of Fig. 2.6.2 before repair (*Ao* aorta, *PA* pulmonary artery, *RV* right ventricle), **b** intraoperative transesophageal echocardiography with color Doppler, showing the left-to-right shunt through the aortopulmonary window, relatively reduced because of the presence of systemic pulmonary hypertension, **c** intraoperative photograph of the same patient showing the external appearance of the aortopulmonary window (white arrows); note the very dilated right ventricle and the two elastic vessel loops (orange color) prepared around the right and left pulmonary artery branches (*RA* right atrium), **d** after incision of the anterior aspect of the defect (transdefect approach), showing the internal appearance of the large aortopulmonary window; note that the right and left pulmonary arteries are now snared by the elastic vessel loops and the two pump suckers are both introduced into the pulmonary artery







(*Aoc* aortic cannula, *AoCC* aortic cross clamp, *RAc* right atrial cannula), **e** the fist stitch anchoring the PTFE patch to the posterior rim of the aortopulmonary window, **f** the progression of the suture of the PTFE patch to the posterior rim of the aortopulmonary window, **g** the progression of the inferior portion of the suture of the PTFE patch to the posterior rim of the aortopulmonary window to reach the arterial wall, **h** the first stitch taking the wall of both great arteries to sandwich the anterior portion of the PTFE patch between them, **i** the progression of the suture sandwiching the anterior portion of the PTFE patch between the aortic and pulmonary artery anterior wall, **j** the progression of the inferior portion of the sandwiching suture of the anterior portion of the PTFE patch between the aortic and pulmonary artery anterior wall, **k** the beginning of the superior portion of the sandwiching suture of the anterior portion of the PTFE patch between the aortic and pulmonary artery anterior wall, **l** the last untied stitches of the sandwiching suture of the anterior portion of the PTFE patch between aortic and pulmonary artery anterior wall, **m** intraoperative transesophageal echocardiography of the same patient with color Doppler, showing complete disappearance of the left-to-right shunt after patch closure of the aortopulmonary window, **n** postoperative echocardiography of the same patient with parasternal short axis view, showing the patch closure (white arrow) of the aortopulmonary window (**n** = photograph courtesy of Dr. Stefano di Bernardo)

fect, or to fatal bleeding, due to the very thin and fragile wall of the window. Now this surgical approach is reserved only for the cases with aortopulmonary window with a morphology "ductus-like", with relatively small diameter and long shape, allowing for complete dissection and control without cardiopulmonary bypass, and without distortion of the origin of a coronary artery.

Closure on cardiopulmonary bypass: aortic cannulation must take into account the position of the aortopulmonary window; therefore, it must be performed as far downstream as possible. Immediately after the beginning of cardiopulmonary bypass, the right and left pulmonary artery must be controlled with tourniquets and snared. The position of the aortic cross also clamp must take into account the location of the pulmonary window. The surgical approach can be transaortic, transpulmonary, transdefect (Fig. 2.6.5) or combined transaortic and transpulmonary (Fig. 2.6.6), according to the morphology of the defect and the personal experience of the surgeon. In any case, after initial opening of the aortic side (Fig. 2.6.7) or of the anterior wall of the defect, the origin of the coronary arteries and the origin of the right pulmonary artery must be very well identified. Therefore, the transpulmonary approach is not ideal, because it does not allow adequate visualization of the origin of the coronary arteries.

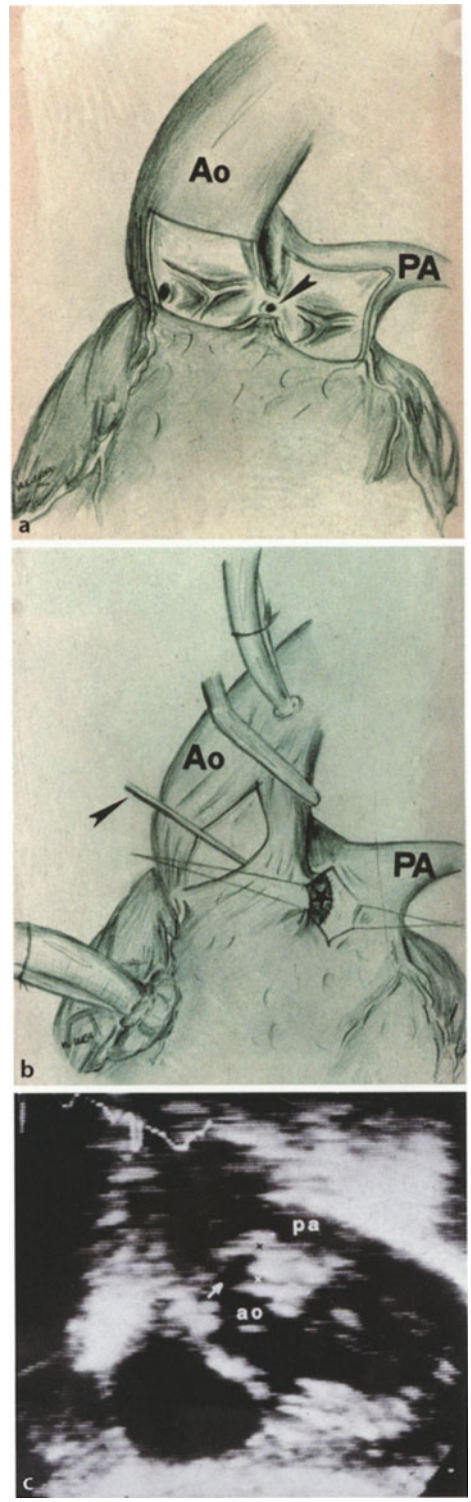


Fig. 2.6.6. Aortopulmonary window: surgery. **a** Artist's view of the preoperative anatomy of the child in Fig. 2.6.3, with the ductus-like aortopulmonary window with anomalous origin of the left coronary artery (black arrow) (*Ao* aorta, *PA* pulmonary artery), **b** Artist's view of the anatomy of the same child with a probe (black arrow) in situ in the anomalous left coronary artery through an aortic incision, patch (asterisk) closure of the pulmonary artery side of the defect, **c** postoperative echocardiography of the same patient showing the same image of Fig. 2.6.3, with the patch (black asterisk) closing the pulmonary artery side of the defect, leaving the anomalous origin of the left coronary artery (white asterisk) on the aortic side of the defect (reproduced with permission from Corno AF, Pierli C, Biagioli B, Lisi G, Grossi A (1988) Anomalous origin of the left coronary artery from an aorto-pulmonary window. J Thorac Cardiovasc Surg 96:669–671)

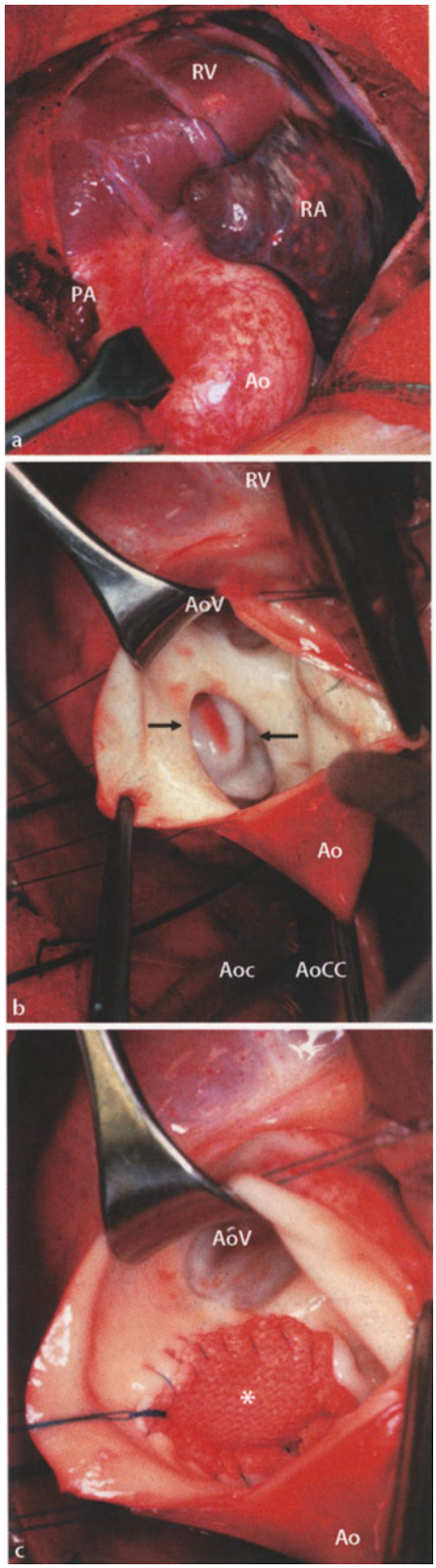


Fig. 2.6.7. Aortopulmonary window: surgery. **a** Intraoperative photograph of the surgical repair with the external appearance of a large aortopulmonary window (*Ao* aorta, *RA* right atrium, *RV* right ventricle, *PA* pulmonary artery), **b** the surgical repair in the same patient with the large aortopulmonary window (black arrows) exposed through an aortotomy (*Aoc* aortic cannula, *AoCC* aortic cross clamp, *AoV* aortic valve), **c** the completed surgical repair in the same patient with the patch (white asterisk) closure of the aortopulmonary window

Patch closure with PTFE, Dacron or polyester patch, is always preferred to the direct closure, to avoid distortion of the surrounding structures, particularly of the origin of the coronary arteries.

In the presence of anomalous origin of a coronary artery, tunnel or coronary artery reimplantation (like for the arterial switch operation, see the chapter "Transposition of the great arteries") are required in order to leave the origin of the anomalous coronary artery in connection with the aortic side of the circulation (Fig. 2.6.6).

Potential complications

Residual or recurrent left-to-right shunt, residual or progressive obstruction of the ascending aorta or the main pulmonary artery, obstruction to a coronary artery origin.

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Chapter 2.7 Anomalous pulmonary arteries

In this chapter only the anomalous origin of a pulmonary artery from the systemic circulation, without interposition of ductal tissue, is taken in consideration. The congenital vascular anomaly in which the left pulmonary artery originates from the right pulmonary artery or pulmonary artery sling is treated separately (see chapter "Slings and rings"). The origin of one or both pulmonary arteries from the transverse aortic arch via a ductus arteriosus or collateral arteries, or from the descending thoracic aorta via collateral arteries, is treated separately (see chapter "Pulmonary atresia with ventricular septal defect").

Incidence

Isolated origin of a pulmonary artery from the systemic circulation is an extremely rare congenital malformation, present in only 20% of patients with anomalous origin of a pulmonary artery. Fewer than 200 cases have been so far reported in the literature.

Morphology

The anomalous origin of the pulmonary artery (called also hemitruncus) is a rare congenital malformation in which only one pulmonary artery branch, usually the right (opposite to the laterality of the aortic arch), originates from the posterior aspect of the ascending aorta just above the aortic sinotubular junction, without any defect between the ascending aorta and the main pulmonary artery, whereas the main pulmonary artery and the other pulmonary artery branch arise in their normal position, therefore are connected with the morphological right ventricle. The other main difference with the persistent truncus arteriosus is the presence of two well-separated semilunar valves, aortic and pulmonary valves, and the usual absence of ventricular septal defect. Therefore hemitruncus defines only the anomalous origin of one pulmonary artery. The anomalous pulmonary artery, most frequently the right, has unrestricted origin, and normal structure, course and distribution. The anomalous origin of the left pulmonary artery (isolated lesion in 40% of patients) usually occurs in the presence of right aortic arch. Very rarely the anomalous pulmonary artery can take origin from the descending thoracic aorta or from either the right or the left coronary artery. Extremely rare cases have been reported of anomalous origin of one pulmonary artery from the innominate artery, the right pulmonary artery with normal left aortic arch, and the left pulmonary artery in the presence of right aortic arch.

Associated anomalies

Left (rarely right) patent ductus arteriosus is present in 50–75% of patients. Other associated anomalies are atrial or ventricular septal defect (8–10% of patients), tetralogy of Fallot with or without absent pulmonary valve, pulmonary stenosis or atresia with ventricular septal defect, aortopulmonary window, right aortic arch (reported in 50–75% of cases with

A. F. Corno, *Congenital Heart Defects*

anomalous left pulmonary artery), aortic coarctation, aortic arch interruption.

Pathophysiology

The typical pathophysiolgic pattern is a large left-to-right shunt, like in large patent arteriosus, with distribution only to one lung, exposed to systemic pressure and oxygen saturation. The consequence is unilateral ventilation-perfusion mismatch in the lung perfused by the anomalous pulmonary artery, and systemic pressure in the anomalous pulmonary artery. On the other side, the contralateral lung, receiving the entire right ventricular output, can over time develop pulmonary hypertension, but still reactive to oxygen inhalation and with reversible vascular lesions, as demonstrated in our experimental model of unilateral pulmonary hypertension.

Diagnosis

Clinical pattern: dyspnea, respiratory distress, hemoptysis; bounding peripheral pulses; cardiac murmur can be only systolic or even absent; cyanosis can be a consequence of the ventilation-perfusion mismatch of the affected lung.

Electrocardiogram: biventricular and right atrial enlargement.

Chest X-ray: cardiomegaly, with heart assuming a globular shape; dishomogeneous pulmonary vascularity.

Echocardiogram: echocardiography is the method for diagnosis; the subxiphoid right oblique projection (Fig. 2.7.1) shows the presence of two concordant ventricular outflow tracts, absence of the usual main pulmonary artery bifurcation pattern and the right or left anomalous pulmonary artery arising directly from the aorta, with the main pulmonary artery continuing with the controlateral pulmonary branch; all the needed information on the position of the anomalous pulmonary artery is provided; Doppler color flow allows indirect measurement of the anomalous pulmonary artery pressure.

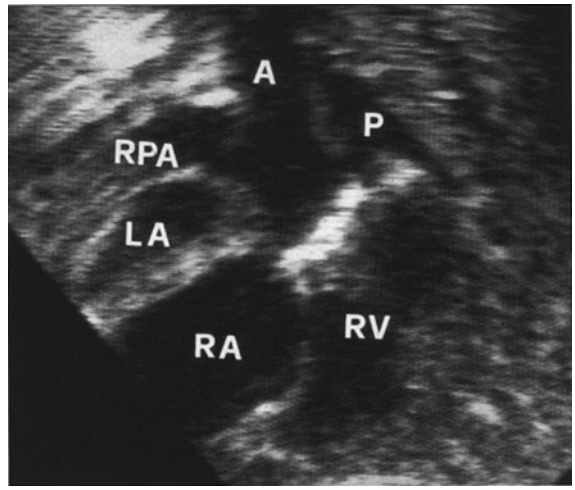

Fig. 2.7.1. Anomalous pulmonary arteries: echocardiography. Subxiphoid right oblique projection showing the absent connection of the right pulmonary artery with the main pulmonary artery and the anomalous origin of the right pulmonary artery directly from the aorta (*A* aorta, *LA* left atrium, *P* main pulmonary artery, *RA* right atrium, *RPA* right pulmonary artery, *RV* right ventricle) (reproduced with permission from Marino B, Thiene G (1990) Atlante di anatomia ecocardiografica delle cardiopatie congenite, USES, Firenze)

Cardiac catheterization: useful to confirm the echocardiographic diagnosis (Fig. 2.7.2), to rule out associated lesions and to calculate the value of pulmonary vascular resistance, particularly in older children coming to observation after late referral.

Indications for surgical treatment

Surgery is indicated to prevent the development of monolateral pulmonary vascular obstructive disease in the lung perfused from the anomalous pulmonary artery with high pressure, high flow and elevated oxygen saturation, as demonstrated in our experimental model of unilateral pulmonary hypertension. High mortality (80% by the age of one year) has been reported in patients not undergoing surgery; the advantages of prenatal

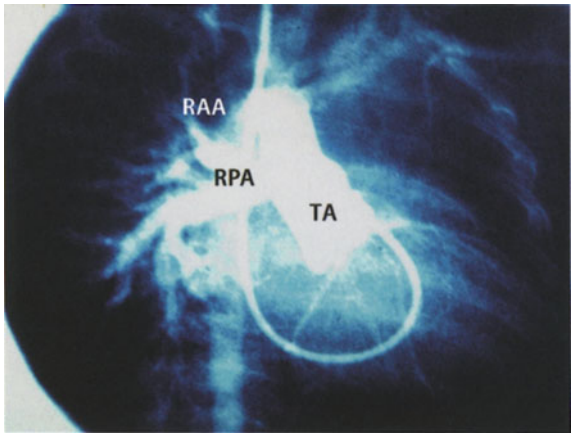

Fig. 2.7.2. Anomalous pulmonary arteries: angiography. Anteroposterior view of a contrast injection in the root of a truncus arteriosus, giving origin to the aorta with right aortic arch, right pulmonary artery and coronary arteries; the left pulmonary artery is not visualized (*RAA* right aortic arch, *RPA* right pulmonary artery, *TA* truncus arteriosus)

diagnosis have been demonstrated in this type of malformation, referred to surgery in the neonatal period, before the development of vascular lesions.

Surgical treatment

The surgical repair can be performed either with or without cardiopulmonary bypass. The use of cardiopulmonary bypass, particularly in infants, allows adequate mobilization of the anomalous pulmonary artery, safe separation from the ascending aorta and reimplantation on the main pulmonary artery. Of course aortic cannulation must take into account the position of the anomalous pulmonary artery; thus, it must be performed as far downstream as possible. Immediately after the beginning of cardiopulmonary bypass, the anomalous pulmonary artery must be snared. If the surgical technique requires aortic cross clamping, the position of the aortic cross clamp also must take into account the location of the anomalous pulmonary artery. The anomalous pulmonary artery is then divided at its origin from the aorta, and the remaining defect is closed with a patch (PTFE, Dacron).

Various surgical techniques have been reported for reimplantation of the anomalous pulmonary artery to the main pulmonary artery, posterior to the ascending aorta, including the direct anastomosis (definitely the preferred technique, Figs. 2.7.3 and 2.7.4), an extension with either an aortic flap or an autologous pericardial patch, or the

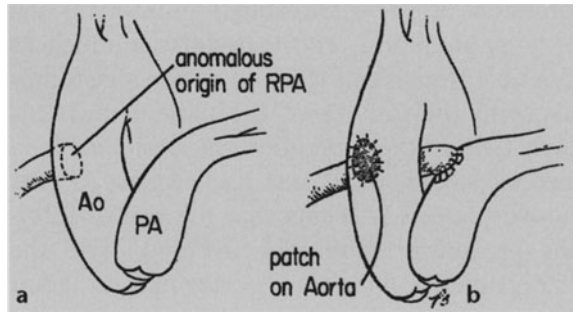

Fig. 2.7.3. Anomalous pulmonary arteries: surgery. Repair: **a** disconnection of the anomalous right pulmonary artery from the aorta, **b** patch closure of the remaining aortic opening, and direct reimplantation of the right pulmonary artery to create the pulmonary artery bifurcation, posteriorly to the aorta (reproduced with permission from Richardson JV, Doty DB, Rossi NP, Ehrenhaft JL (1979) The spectrum of anomalies of aortopulmonary septation. J Thorac Cardiovasc Surg 78:21–27)

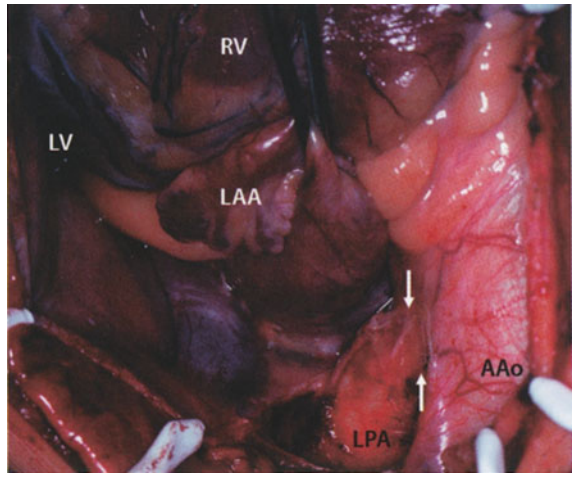

Fig. 2.7.4. Anomalous pulmonary arteries: surgery. Intraoperative photograph showing the anomalous origin (white arrows) of the left pulmonary artery from left lateral aspect of the ascending aorta (*AAo* ascending aorta, *LAA* left auricular appendage, *LPA* left pulmonary artery, *LV* left ventricle, *RV* right ventricle)

interposition of a synthetic tubular prosthesis. The final technique (synthetic tubular prosthesis), the first one historically utilized for the surgical treatment of the anomalous origin of a pulmonary artery, has been practically abandoned, and it should be avoided, particularly in small children,

Whichever the surgical technique utilized, the key points are to avoid tension at the anastomotic site, to avoid the potential compression of the reimplanted pulmonary artery by the aorta, and to reduce as much as possible the risk of stenosis at the anastomosis with the growth of the patient, particularly when the operation is performed in neonates and small infants. Because of the above reasons it seems that the use of tubular prosthesis should be avoided, with the exception of operation performed in adult patients.

Potential complications

In the immediate postoperative period, crises of pulmonary hypertension have been reported, while in the later follow-up residual or recurrent stenosis at the site of the anastomosis with the main pulmonary artery as well as dishomogeneous lung perfusion at the control with scintigraphy have been reported.

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Chapter 2.8 Anomalous coronary arteries

Incidence

Congenital coronary artery anomalies have been estimated to occur in 0.2 to 1.2% of the general population. Since these data are coming from large series of angiographic studies, the incidence is most probably underestimated, since many people never undergo angiography. Among the different types of anomalous coronary artery, the anomalous origin of the left coronary artery from the pulmonary artery is the most common, with an incidence of 0.25–0.50% in congenital heart defects, a little more frequent than coronary artery fistula, found with an incidence of 0.20–0.40% of congenital heart defects.

Morphology

Regardless of the position of the heart within the chest and the position and the origin of the great arteries, aortic and pulmonary valves normally have a single point of contact, with commissural apposition; coronary arteries almost always originate normally from the facing sinuses of Valsalva on either side of this point of commissural contact; coronary arteries do not normally originate from the nonfacing or most distant sinus. However, variations in coronary anatomy occur, and different types of coronary artery anomalies exist:

Isolated anomalous origin of a coronary artery:

- *From the pulmonary artery (historically called Bland-White-Garland syndrome):* in most patients the anomalous left coronary artery (either the left main coronary artery, or the anterior descending or the circumflex coronary artery) originates from the posterior or left facing sinus of the pulmonary artery, very rarely from the right or anterior facing sinus of the pulmonary artery, and exceptionally from the nonfacing sinus; the branching of the anomalous left coronary artery is usually normal; the origin of the right coronary artery is generally normal, but this vessel is usually enlarged and tortuous.
- *From the wrong sinus of Valsalva:* there are several variants of anomalous origin of a coronary artery from the opposite facing sinus of Valsalva, either the right coronary artery originating from the left anterior sinus, or the main trunk of the left coronary artery or one of the two main branches (left anterior descending or circumflex coronary artery) originating from the right anterior sinus or from the right coronary artery; in all these conditions, the course of the coronary artery with anomalous origin can be also anomalous, like being retroaortic, anterior to the right ventricular outflow tract, or between aorta and pulmonary artery; the last is the most dangerous condition, frequently (57 to 64% of the cases, up to 82% with the course of the left main coronary artery between the great arteries) associated with sudden death.

A. F. Corno, *Congenital Heart Defects*

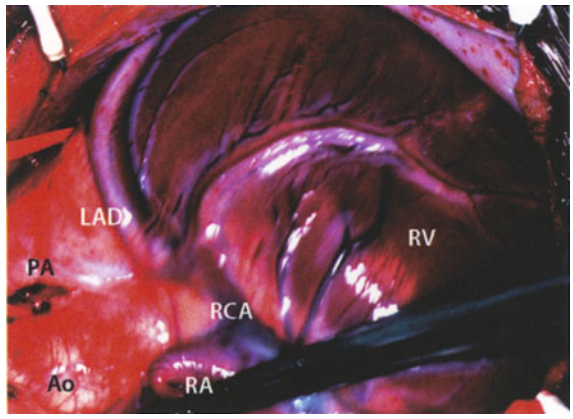

Fig. 2.8.1. Anomalous coronary arteries: morphology. Intraoperative photograph showing an anomalous origin of the left anterior descending coronary artery from the right coronary artery (*Ao* aorta, *LAD* left anterior descending coronary artery, *PA* pulmonary artery, *RA* right atrium, *RCA* right coronary artery, *RV* right ventricle)

- *From another coronary artery:* in this cases there might be a single origin for the coronary arteries from the aortic root, with an anomalous right coronary artery originating from the left anterior descending coronary artery, or still a double orifice for the coronary artery, but with an anomalous left anterior descending coronary artery originating from the right coronary artery and the left facing sinus giving only origin to the circumflex coronary artery (Fig. 2.8.1).
- *From an additional coronary ostium:* normally there are two coronary artery ostia; nevertheless, sometimes there are three coronary artery ostia, because either the conal branch of the right coronary artery originates separately from the right sinus of Valsalva, or because the left anterior descending and circumflex coronary arteries arise separately from the left sinus instead of from the main common coronary trunk.

Anomalous course of a coronary artery:
- normally coronary arteries originate more or less perpendicular to the aortic wall, with the ostia located in the middle of the sinus of Valsalva, just above the free margin of the aortic leaflet and below the sinotubular junction; coronary arteries with ectopical origin usually course tangentially to the aortic wall.
- normally the course of coronary arteries is epicardial; the proximal and sometimes the middle segment of the left anterior descending coronary artery may have an intramural or subepicardial course in 5–25% of the population, but this is not considered an anomaly.

Atresia of a coronary artery orifice: several variations are possible, although very rare:
- atresia of the origin of the main trunk of the left coronary artery,
- atresia or the origin of the left anterior descending coronary artery,
- atresia of the origin of the circumflex artery,
- atresia of the origin of the right coronary artery.

Coronary artery fistula: a coronary artery fistula consists of a single or multiple sizable communication(s) between a normally distributed coronary artery, or its branches, and either a cardiac cavity (=coronary-cameral fistula) or any segment of the systemic or pulmonary circulation (=coronary arteriovenous fistula) bypassing the myocardial capillary phase; coronary artery fistula from either the right (60% of the cases), or the left coronary artery (35%) or both coronary arteries (5%) is in 90–92% of patients connected to the right side of the heart, but it may be connected to:
- pulmonary artery; reported in about 10% of patients with pulmonary atresia with ventricular septal defect, in all cases joining central pulmonary artery (see chapter "Pulmonary atresia with ventricular septal defect"); if a coronary artery arises directly from the pulmonary artery without direct aortic connection, this anomaly is classified as anomalous origin of a coronary artery from the pulmonary artery (see above),
- right ventricle,
- right atrium,
- coronary sinus,

- superior vena cava,
- pulmonary vein,
- left atrium,
- left ventricle,
- multiple.

Congenital coronary artery fistulas may be associated (in 20% to 45% of the cases) with a congenital heart defect: atrial septal defect, ventricular septal defect, pulmonary atresia with intact ventricular septum, tetralogy of Fallot, peripheral pulmonary artery stenosis, aortic atresia, aortic coarctation, patent ductus arteriosus.

Anomalous coronary artery associated with a congenital heart defect: coronary anomalies may be associated with several other congenital heart defects, most notably transposition of the great arteries, tetralogy of Fallot and pulmonary atresia; the specific problems due to the presence of an anomalous coronary artery in these malformations have been discussed in the relative chapters.

Associated anomalies

Ventricular septal defect, tetralogy of Fallot, pulmonary atresia, truncus arteriosus, aortopulmonary window, transposition of the great arteries, congenitally corrected transposition of the great arteries (=double discordance), double-chambered right ventricle, aortic valve disease, supravalvular aortic stenosis (with Williams syndrome), hypoplastic left heart syndrome, aortic coarctation.

Particularly important is the potential association of an anomalous origin of the left coronary artery from the pulmonary artery with patent ductus arteriosus (see chapter "Patent ductus arteriosus"): even if this combination is extremely rare, there are anecdoctical reports of death of the patient after simple closure of a ductus arteriosus, particularly in the presence of an hypertensive ductus arteriosus, because of the sudden myocardial ischemia and acute myocardial infarction due to the simultaneous presence of the anomalous coronary artery with origin from the pulmonary artery.

Atresia of a coronary artery orifice is more frequently associated with supravalvular aortic stenosis and with anomalies of the aortic or truncal valve, where excess tissue can obstruct the coronary orifice, or where the angle of origin of the coronary artery produces a valve-like obstruction.

Quadricuspid aortic valves can be associated with single coronary ostium, and displacement of the left or right coronary orifice.

Pathophysiology

While the myocardium has a very limited capacity for anaerobic metabolism, the capacity for oxygen extraction is great, although relatively fixed; therefore the heart has a negligible ability to tolerate periods of ischemia, but limited degrees of hypoxemia are generally well tolerated. However, since the oxygen extraction is more or less fixed, any request for an increase in oxygen demand must be met by an increase in myocardial blood flow. The major regulators of coronary blood flow are intramural pressure, aortic diastolic perfusion pressure, myocardial metabolic rate (dependent upon heart rate, inotropic state and systolic arterial pressure), autonomic nervous system control, endothelial function and blood viscosity in response to decreased myocardial oxygen supply. Myocardial ischemia is the result of an imbalance of the demand/supply ratio of oxygen, either for congenital or acquired coronary artery disease.

Anomalous origin of a coronary artery from the pulmonary artery: the pathophysiology is highly variable and depends on age of the patient, rapidity of closure of the patent ductus arteriosus, pulmonary artery pressures and resistance, presence and development of intercoronary collateral arteries between the right and left coronary artery systems providing retrograde perfusion of the anomalous left coronary artery, and de-

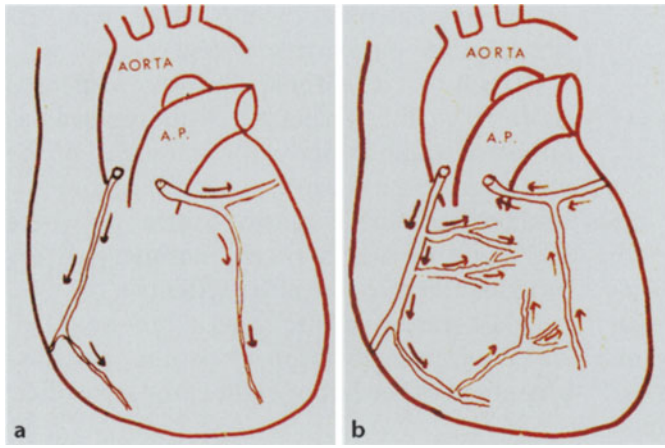

Fig. 2.8.2. Anomalous coronary arteries: pathophysiology. **a** Anomalous left coronary artery from the pulmonary artery: stage 1, **b** anomalous left coronary artery from the pulmonary artery: stage 2

gree of myocardial ischemia. Four pathophysiologic stages have been described:

- Stage 1 (Fig. 2.8.2 a): in the neonatal period the pulmonary vascular resistance is high, with systemic pulmonary artery pressure; the perfusion of the anomalous left coronary artery is adequate, despite the low oxygen saturation, without signs of myocardial ischemia or left ventricular dysfunction;
- Stage 2 (Fig. 2.8.2 b): with the physiologic decrease of pulmonary vascular resistance and the subsequent decrease of pulmonary artery pressure, the perfusion of the anomalous left coronary artery becomes inadequate, and collateral flow develops from the right coronary artery system; this collateral flow from the right coronary artery system meets the high resistance of the left ventricular myocardium, and preferential retrograde blood flow occurs through the anomalous left coronary artery towards the low pressure pulmonary artery (constituting a left-to-right shunt), with signs of myocardial ischemia or infarction; a certain degree of left-to-right shunt develops, with a subsequent coronary arterial steal;
- Stage 3: rarely (10 to 15% of the patients) a very large collateral circulation develops between the right and left coronary artery systems, able to maintain adequate coronary artery perfusion with adequate perfusion of the anomalous left coronary artery, with mild myocardial damage, sometimes through adulthood;
- Stage 4: eventually, the collateral coronary blood flow becomes inadequate, with retrograde flow in the anomalous left coronary artery, and myocardial ischemia or infarction occurs.

Myocardial ischemia typically occurs in the anterolateral left ventricular myocardium, causing global left ventricular dilatation and dysfunction. Mitral valve regurgitation is quite common, secondary either to ischemia or infarction of a papillary muscle and/or to dilatation of the valve annulus and/or to extensive left ventricular fibrosis. Left atrial dilatation and pulmonary venous congestion (subsequent to both the left-to-right shunt and to the left heart failure) follow, adding the signs of congestive heart failure or cardiogenic shock to those of angina pectoris.

Coronary artery fistula: the pathophysiologic mechanism is a myocardial stealing due to the reduction of myocardial blood flow distal to the site of the coronary artery fistula connection. The mechanism is related to the variables of diastolic pressure gradient and run-off from the coronary circulation to a low-pressure receiving cavity or vessel; the intracoronary perfusion pressure progressively diminishes accordingly with the size of the fistula, and therefore with the degree of left-to-right shunt. The compensation occurs by

means of a progressive enlargement of the origin and course of the feeding coronary artery. Eventually the myocardium distal to the site of the fistula is at risk for ischemia, most frequently becoming evident with increased myocardial oxygen demand, e.g. during physical exercise. With time the coronary artery leading to the fistula progressively dilates, which, in turn, may progress to aneurysm formation, intimal ulceration, medial degeneration, intimal rupture, atherosclerosis, calcification, side-branch obstruction, mural thrombosis, and, more rarely, rupture.

Diagnosis

Clinical pattern: this depends upon the type of congenital coronary artery anomaly; gallop rhythm (=third heart sound), holosystolic cardiac murmur of mitral regurgitation, continuous murmur (in the presence of coronary artery fistula); respiratory distress, feeding intolerance, dyspnea (at rest or on effort), tachypnea, tachycardia, fatigue, palpitations, atypical angina, myocardial ischemia or infarction, pulmonary edema; in infants angina is recognized by the presence of irritability, diaphoresis, poor feeding, failure to thrive, wheezing, sweating, tachypnea, poor peripheral perfusion (pallor, ashen gray color) due to low cardiac output; unfortunately in a substantial number of patients the symptoms may not be evident before syncope or sudden death occurring after exertion, particularly in adolescents or young adults.

Electrocardiogram: arrhythmias, atrial fibrillation, ventricular tachycardia, signs of myocardial infarction (Q waves and ST-segment elevation in anterolateral leads) or myocardial ischemia (ST elevation, T wave inversion), left atrial and ventricular enlargement (in the presence of coronary artery fistula).

Chest X-ray: cardiomegaly, left atrial and ventricular enlargement, pulmonary venous congestion, interstitial edema.

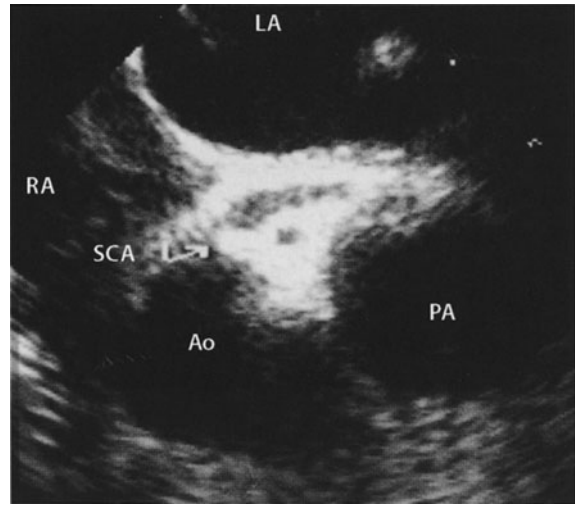

Fig. 2.8.3. Anomalous coronary arteries: echocardiography. Echocardiography showing the single coronary artery origin (indicated by a white arrow) from the aorta (*Ao* aorta, *LA* left atrium, *PA* pulmonary artery, *RA* right atrium, *SCA* single coronary artery) (photograph courtesy of Dr. Nicole Sekarski)

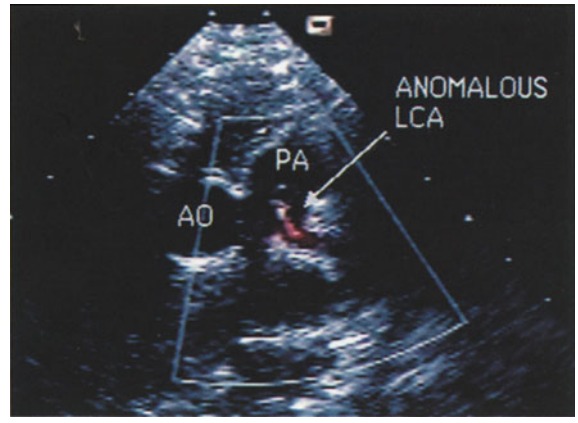

Fig. 2.8.4. Anomalous coronary arteries: echocardiography. Anomalous left coronary artery from the pulmonary artery. Parasternal short-axis view showing the anomalous origin from the pulmonary vartery with retrograde flow (*Ao* aorta, *LCA* left coronary artery, *PA* pulmonary artery) (photograph courtesy of Dr. Nicole Sekarski)

Echocardiogram: visualization of the anomalous coronary artery is possible with two-dimensional echocardiography (Fig. 2.8.3), Doppler color flow shows anomalous retrograde diastolic flow in the left coronary artery (constituting a left-to-right shunt) originating from the pulmonary artery (Fig. 2.8.4); dilated left ventricular cavity with poor function, re-

duced ejection fraction, reduced fractional shortening, mitral regurgitation; left atrial and ventricular enlargement due to volume overload in the presence of coronary artery fistula.

Cardiac catheterization: indicated in the presence of anatomy of the coronary arteries not fully defined by echocardiography; aortic root injection may be sufficient to accurately define a specific coronary artery anomaly, particularly regarding the origin and the distribution of the coronary arteries (Figs. 2.8.5 and 2.8.6), but selective coronary artery studies are sometimes necessary; in the presence of anomalous left coronary artery from the pulmonary artery, the delayed passage of contrast medium and the typical blush from the anomalous left coronary artery into the main pulmonary artery can be observed (Fig. 2.8.7); transcatheter coil or device embolization of a coronary artery fistula (Fig. 2.8.8) may be performed during cardiac catheterization.

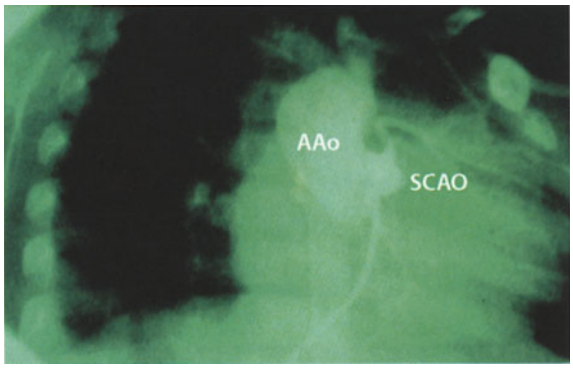

Fig. 2.8.5. Anomalous coronary arteries: angiocardiography. Contrast injection in the ascending aorta showing the single origin of the coronary arteries (*AAO* ascending aorta, *SCAO* single coronary artery origin)

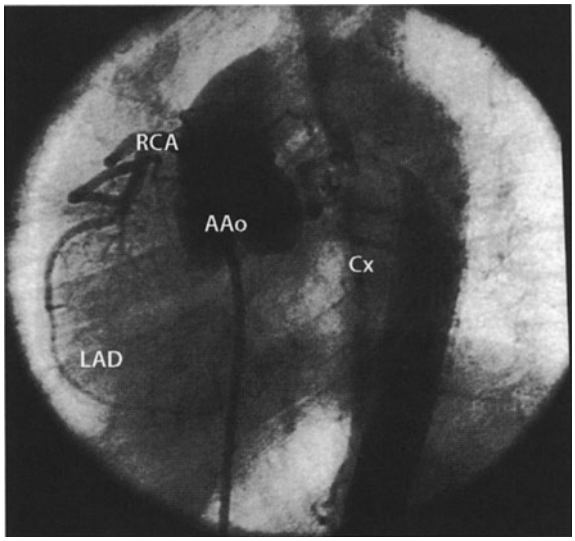

Fig. 2.8.6. Anomalous coronary arteries: angiocardiography. Contrast injection in the ascending aorta showing the anomalous origin of the left anterior descending coronary artery from the right coronary artery (*AAo* ascending aorta, *CxA* circumflex coronary artery, *DTAo* descending thoracic aorta, *LAD* left anterior descending coronary artery, *RCA* right coronary artery)

Magnetic resonance: since the incidence of incorrect diagnosis with conventional coronary angiography is reported to be as high as 50% in children with anomalous coronary arteries, fast magnetic resonance angiography has become the gold standard diagnostic procedure in these patients; transverse sections provide precise details on the origin and proximal course in relationship with the position of the great arteries.

Indications for surgical treatment

An infant with symptoms of myocardial ischemia due to an anomalous coronary artery requires intensive treatment with initial efforts to reduce the myocardial oxygen demand followed by a rapid decision-making process.

The presence of an anomalous coronary artery is a potentially life-threatening condition, based on the anatomy, the development of collateral circulation, and the presence and the type of associated lesions. In particular two types of anomalous coronary artery present with a very poor natural history:

- the anomalous origin of the left coronary artery from the pulmonary artery: nearly 90% of the untreated patients die within the first year of life;
- the anomalous origin of a coronary artery from the wrong aortic sinus, particularly with course of a coronary artery branch

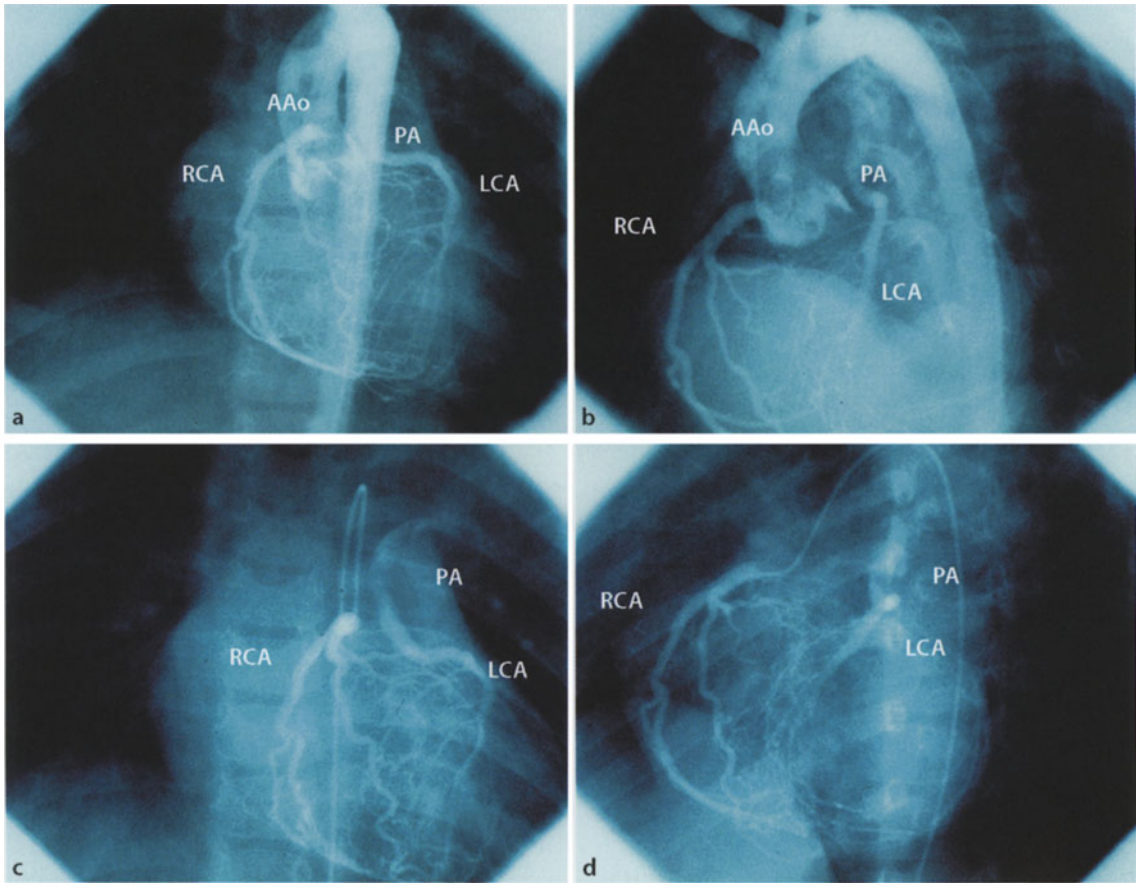

Fig. 2.8.7. Anomalous coronary arteries: angiocardiography. Anomalous left coronary artery from the pulmonary artery. **a** Anteroposterior view and **b** left anterior oblique view with contrast injection in the ascending aorta and (*AAo* ascending aorta, *LCA* left coronary artery, *PA* pulmonary artery, *RCA* right coronary artery), **c** anteroposterior view and **d** left anterior oblique view with selective contrast injection in the right coronary artery showing the opacification of the right coronary artery, the retrograde opacification of the left coronary artery, not originating from the aorta, through a very well-developed collateral circulation coming from the right coronary artery, and the initial opacification of the internal wall of the pulmonary artery

between the two great arteries: these patients are exposed to the risk of sudden death.

Myocardial ischemia and infarction, global cardiomyopathy, chronic mitral regurgitation, congestive heart failure and sudden death are the major risks in the natural history of anomalous coronary arteries.

Mitral regurgitation, sometimes present before surgery due to the left ventricular ischemic dysfunction as a consequence of the anomalous coronary arteries perfusion, can be substantially reduced by the relief of myocardial ischemia and the reconstruction of a two coronary artery system, with subsequent myocardial recovery; therefore simultaneous mitral valve surgery seems unwarranted. Nevertheless, in the presence of severe mitral valve regurgitation in older children or adolescents due to irreversible myocardial injury or papillary muscle infarction, mitral valve repair at the time of coronary artery reimplantation may be required.

Nevertheless, different types of surgical procedures have been used in the past to treat an anomalous coronary artery, the experience gained in coronary artery transfer (=mobilization and reimplantation into the new aortic root) during the arterial switch

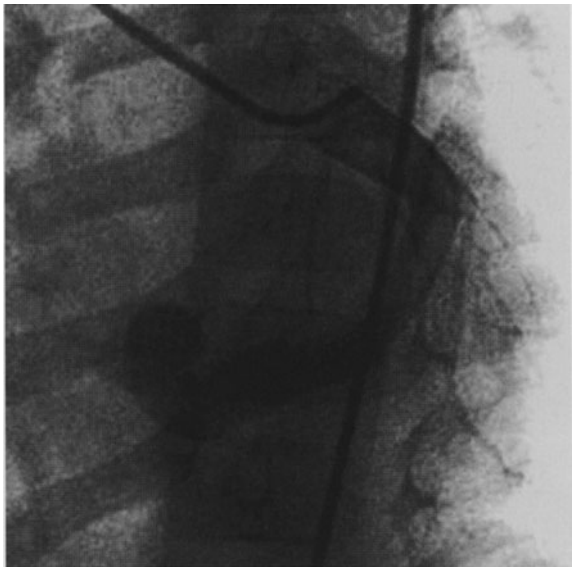

Fig. 2.8.8. Anomalous coronary arteries: angiography with left anterior oblique view showing a coronary artery fistula to the right ventricle (photograph courtesy of Dr. Shakeel Qureshi)

for transposition of the great arteries (see chapter "Transposition of the great arteries") reduced the perioperative risk of coronary artery reimplantation enough to propose this type of surgical approach as the best form of treatment for anomalous coronary artery, in order to reconstruct a two coronary artery system and obtain the best physiological repair with myocardial recovery.

Spontaneous closure may rarely occur in very small coronary artery fistulas; therefore, there is generally indication for closure, with the exception of very small shunt (QP/QS < 1.3). Atresia of a coronary artery orifice and presence of an intramural course require individualized treatment.

Surgical treatment

Anomalous origin of the left coronary artery from the pulmonary artery:

- Ligation: the simple ligation of the origin of the anomalous coronary artery, performed without cardiopulmonary bypass through a median sternotomy or from a limited left anterior thoracotomy, has been used quite extensively in the past with acceptable survival rate, particularly in the presence of well developed collateral circulation. The follow-up of children operated on with this technique was nevertheless complicated by suboptimal left ventricular performance, and the technique has been practically abandoned in favor of the reimplantation of the anomalous coronary artery to the aorta.
- End-to-side anastomosis of the left subclavian artery or the internal mammary artery (= thoracic artery) to the anomalous coronary artery: these techniques, also not requiring cardiopulmonary bypass, provide an arterial revascularization of the anomalous coronary artery by end-to-side anastomosis of the transected left subclavian artery or the internal mammary artery, but both have been nearly completely abandoned. The only indication to use these techniques is remote distance of the anomalous coronary artery ostium within the pulmonary artery from the adjacent aorta, making direct coronary transfer impossible and the Takeuchi procedure difficult.
- Takeuchi procedure: for children with anatomy unsuitable for the anomalous coronary artery reimplantation, or when the surgeon is not comfortable with the surgical technique of aortic reimplantation, an alternative surgical technique exists, nowadays rarely utilized. An aortopulmonary window is created, on cardiopulmonary bypass, carefully avoiding injury to the aortic valve, then an intrapulmonary artery tunnel (= *intrapulmonary baffling technique*) connecting the created aortopulmonary window with the anomalous coronary artery origin is obtained by opening the pulmonary artery and creating an anterior transverse flap of the native pulmonary artery wall or a pericardial or PTFE patch. The pulmonary artery is then repaired with direct suture with a patch of autologous or heterologous pericardium.

An alternative technique is to use elongated flaps of both the aortic and pulmonary artery wall, sewn side to side, to create

a tunnel from the aorta to the origin of the anomalous left coronary artery.

- Reimplantation (Fig. 2.8.9): with cardiopulmonary bypass (even if a case has been reported in an adult patient with a minimally invasive approach, without cardiopulmonary bypass). After accurate identification of the anatomy, the pulmonary artery is transected, and the anomalous left coronary artery is carefully mobilized and removed with a button of tissue around the orifice with a surgical technique similar to the technique used for the arterial switch operation (see chapter "Transposition of the great arteries"). After an incision in the ascending aorta (with or without removal of a small button of aortic wall) in accord with the identified position for coronary artery reimplantation, the anomalous left coronary artery, turned posteriorly, is directly anastomosed into the aortic root. Direct aor-

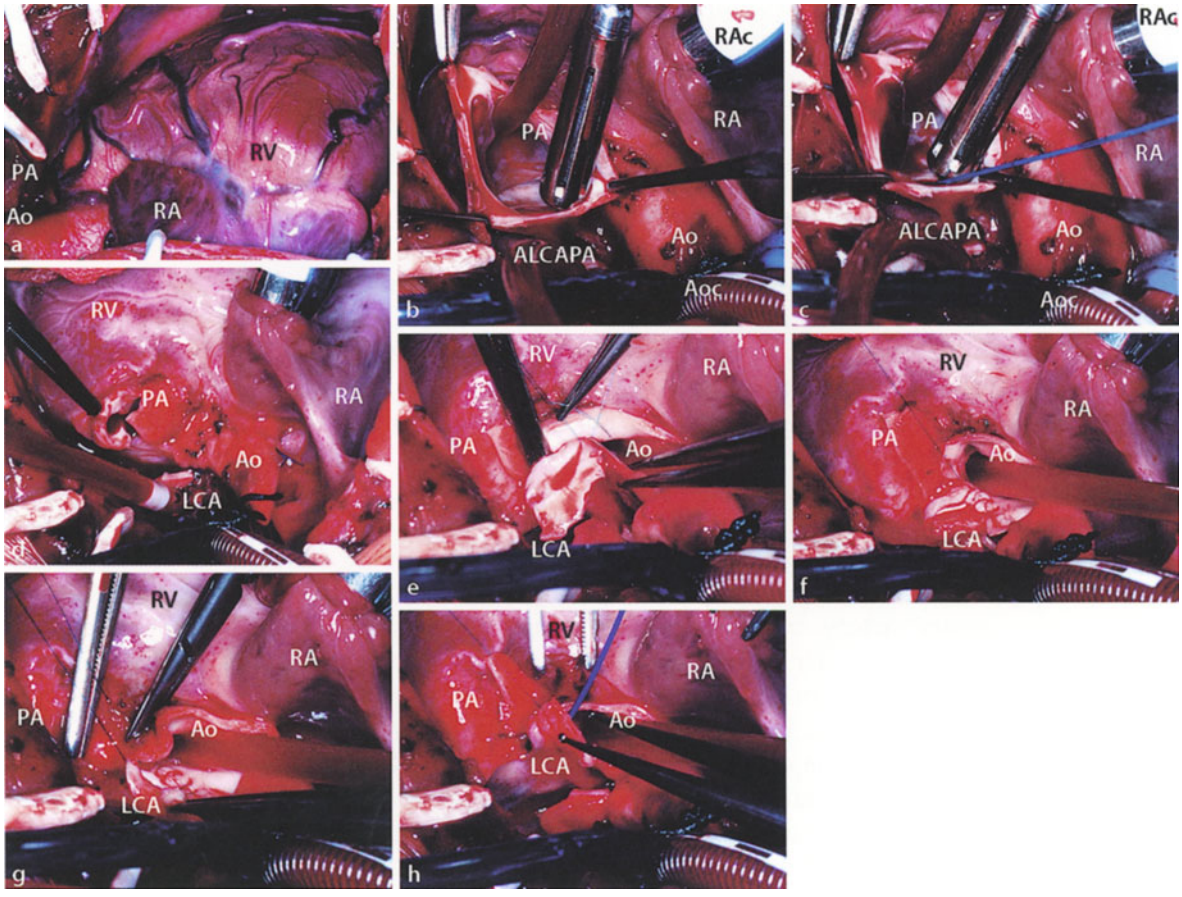

Fig. 2.8.9. Anomalous coronary arteries: surgery. **a** Intraoperative photograph of the same patient of Fig. 2.8.4 with anomalous origin of the left coronary artery from the pulmonary artery showing (**a**) the extensive arterial and venous collateral circulation developed on the epicardium and great arteries (*Ao* aorta, *PA* pulmonary artery, *RA* right atrium, *RV* right ventricle), **b** intraoperative photograph showing the anomalous origin of the left coronary artery from the transected pulmonary artery (*ALCAPA* anomalous left coronary artery from the transected pulmonary artery, *Aoc* aortic cannula, *RAc* right atrial cannula), **c** the anomalous origin of the left coronary artery from the transected pulmonary artery, with the orifice indicated by a blue plastic probe, **d** the anomalous origin of the left coronary artery already separated from the transected pulmonary artery (*LCA* left coronary artery), **e** the beginning of the posterior anastomosis for reimplantation of the left coronary artery to the aorta, **f** completed posterior anastomosis of the left coronary artery to the aorta, **g** beginning of the anterior anastomosis of the left coronary artery to the aorta, **h** almost completed anterior anastomosis of the left coronary artery to the aorta, with the orifice shown by a blue plastic probe

tic reimplantation can be performed also with a roll shaped transected main pulmonary arterial or aortic wall graft. The pulmonary artery is then repaired with direct suture with a patch augmentation of autologous or heterologous pericardium.

- Heart transplantation: when the long-lasting consequences of global myocardial ischemia and the presence of severe and irreversible myocardial damage preclude any form of repair or palliation of the anomalous coronary artery, heart transplantation has been and must be taken into consideration as a valid alternative option. In these cases heart transplantation is performed with cardiopulmonary bypass, using the standard surgical technique.

Anomalous origin of a coronary artery from the wrong aortic sinus: the coronary artery with anomalous origin from a wrong sinus, frequently with an intramural segment, is moved to the appropriate sinus without unroofing; when it is necessary, a new coronary ostium is created, without interfering with the aortic valve commissurae; an alternative technique consists of a coronary artery bypass graft, preferably with implantation of the internal mammary artery (=thoracic artery) to the anomalous coronary artery; in the presence of compression of the anomalous coronary artery between aorta and pulmonary artery, a feasible surgical technique consists of the transection of the main pulmonary artery at the level of its bifurcation, patch closure of the distal opening and reimplantation of the main pulmonary artery to the left pulmonary artery branch; this technique allows for separation of the main pulmonary artery from the aorta, with elimination (or at least reduction) of the coronary artery compression.

Coronary artery fistula: through a median sternotomy the feeding artery of the coronary artery fistula, its course and site of insertion are identified; in rare cases where the fistula is a terminal coronary artery, surgical occlusion by means of a direct ligature can be performed without cardiopulmonary bypass; however, in the vast majority of patients the surgical procedure requires cardiopulmonary bypass and includes opening of the chamber where the anomalous fistula is draining, identification of the fistula, and suture of the anomalous coronary connection; large aneurysms may require surgical resection.

Atresia of a coronary artery orifice: angioplasty of the origin of a coronary artery is the surgical technique used to treat the atresia of a coronary artery orifice, or to enlarge the coronary artery main stem in the presence of atresia of the left main coronary artery; the enlargement can be obtained on cardiopulmonary bypass with a patch of autologous saphenous vein or pericardium, or with a PTFE patch.

Potential complications

Anomalous origin of the left coronary artery from the pulmonary artery:

- Takeuchi procedure: potential complication is the residual or recurrent stenosis of the new coronary artery channel, with subsequent myocardial ischemia, as well as baffle leaks, residual or recurrent supravalvular pulmonary stenosis, due to the obstruction created by the space occupied by the new coronary artery channel constructed inside the main pulmonary artery. Another potential complication is injury to the aortic valve when creating the aortopulmonary window.
- Reimplantation: low cardiac output with hemodynamic instability can complicate the postoperative period, particularly in patients with severely compromised myocardial function and/or operated on after late referral; in these cases a period with mechanical assistance (either left ventricular or biventricular assistance, or extracorporeal membrane oxygenation, rarely intraaortic balloon counterpulsation) and delayed sternal closure can be required, particularly if the adequate precautions

for myocardial protection (administration of antegrade blood cardioplegia after snaring the pulmonary arteries, followed by retrograde cardioplegia administration) have not been observed during surgery. Another potential complication is the obstruction of the reimplanted coronary artery, due to excessive tension, kinking or twisting of the coronary artery, with potential subsequent myocardial ischemia or infarction. Ventricular arrhythmias can accompany poor ventricular function and severe ventricular dilatation.

- Angioplasty of the origin of a coronary artery: despite patch enlargement of the orifice of a coronary artery generally providing adequate coronary artery perfusion, residual or recurrent coronary artery stenosis can require re-operation, in most cases with coronary artery bypass graft accomplished with the internal thoracic (= mammary) artery.

Anomalous origin of a coronary artery from the wrong aortic sinus: inadequate coronary artery perfusion can persist after surgery, as well as malfunctioning of the aortic valve, damaged by the coronary artery reimplantation.

Coronary artery fistulas: myocardial ischemia or infarction, recurrence of the coronary artery fistula.

Atresia of a coronary artery orifice: myocardial ischemia or infarction.

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Chapter 2.9 Mitral valve disease

In this Chapter only congenital malformations of the mitral valve are considered, excluding atrioventricular septal defects, which are separately treated (see chapter "Atrioventricular septal defects"), as well as mitral atresia (see chapter "Hypoplastic left heart syndrome").

Incidence

Congenital mitral valve disease is a very rare entity, occurring in 0.21–0.50% of patients with congenital heart defects. No sex prevalence is reported.

Mitral valve stenosis

Morphology

Congenital mitral stenosis can present in several forms, with narrowing of the effective mitral valve orifice at the supravalvular, valvular or subvalvular level, because of the presence of malformations of one or more of the four major components of the mitral valve: the annulus, the leaflets, the chordae tendineae and the papillary muscles.

From the morphological point of view, mitral stenosis, or mitral valve disease with prevalent stenosis (55–60% of the cases), can be due to hypoplasia of the mitral valve annulus, fusion of the commissurae and/or chordae, parachute mitral valve (= valve with all the chordae attached to a single papillary muscle, generally the posterior papillary muscle, with absence of the anterior papillary muscle), thick and short chordae tendineae, fusion between papillary muscle and leaflets, double orifice mitral valve, mitral arcade (= anomalous papillary muscle arcade with a bridge of fibrous tissue through the free aspect of the anterior leaflet), unicuspid mitral valve, hammock valve (mitral valve with fused commissurae and with a central orifice obstructed by numerous intermixed short chordae attached to abnormal hypertrophic papillary muscles). Frequently the mitral stenosis is the result of malformations at multiple levels.

Severe hypoplasia of the mitral valve, or atresia, results in hypoplasia of the left ventricular cavity; when the degree of hypoplasia of the left ventricle is so important that the left ventricle is not able to sustain the systemic cardiac output, this situation is considered part of the spectrum of the hypoplastic left heart syndrome (see chapter "Hypoplastic left heart syndrome"). In this chapter only situations with mitral stenosis allowing enough blood into the left ventricle to sustain the systemic cardiac output are considered. One exception is the very rare presence of hypoplastic mitral valve with hypoplastic annulus and adequate size left ventricle because of the simultaneous presence of an unrestrictive ventricular septal defect.

A. F. Corno, *Congenital Heart Defects*

Mitral valve with stenosis and regurgitation, or with combined lesions, occur in about 15% of the cases

Supravalvular mitral stenosis: congenital supravalvular mitral stenosis (or *supravalvular mitral ring*) is a very rare malformation (there are less than 100 cases reported in the literature since its first description in 1902), characterized by the presence of a shelf-like fibrous diaphragm, abnormal ridge of connective tissue, with variable thickness and extent, with 1 or 2 small orifices, positioned on the atrial side of the mitral valve, very close to it, attached at the level of the annulus or just above the annulus, covering and obstructing the mitral valve. Often circumferential in shape, the supravalvular ring may frequently adhere to the mitral valve leaflets and restrict their movements, or even protrude into the orifice of the mitral valve. Pulmonary veins and the left auricular appendage are all located proximally to the ring. The underlying mitral valve may be functionally normal, but in most patients the mitral valve is anatomically abnormal.

Accessory mitral valve tissue: this is an extremely rare congenital malformation causing associated left ventricular outflow tract obstruction.

Associated anomalies

Typically associated (50% of cases) with parachute mitral valve is the *Shone complex* (multiple level systemic obstructions, with supravalvular mitral ring, parachute mitral valve, subvalvular aortic stenosis, and aortic coarctation with or without aortic arch hypoplasia). Associated lesions reported with mitral stenosis are ventricular septal defect (30% of cases), atrial septal defect, complete atrioventricular septal defect, double outlet right ventricle, complete transposition of the great arteries, double discordance (atrioventricular and ventriculoarterial), single ventricle, straddling tricuspid valve, pulmonary stenosis, patent ductus arteriosus.

Supravalvular mitral stenosis: the supravalvular ring is found in combination with other congenital heart defects in about 90% of the cases: the *Shone complex* is the most frequent, but also atrioventricular septal defect, ventricular septal defect, tetralogy of Fallot and patent ductus arteriosus have been reported; less common associations are persistent left superior vena cava, atrial septal defect, double outlet right ventricle and transposition of the great arteries.

Pathophysiology

Mitral stenosis obstructs the blood inflow to the left ventricle, and is, therefore, accompanied by increased left atrial pressure in direct proportion to the severity of the obstruction. The elevated left atrial pressure, in turn, restricts the pulmonary venous return to the left atrium, increasing the pulmonary venous pressure and subsequently the pulmonary artery and right ventricular pressure. The elevated hydrostatic pressure in the pulmonary capillaries forces fluid into alveoli and interstitial space, causing pulmonary congestion. Congested bronchial veins may obstruct small bronchioles, with an increase in airway resistance as a consequence.

The subsequent compensatory mechanism is pulmonary vasoconstriction. When this is severe and prolonged enough it may induce vascular changes like medial thickening and intimal fibrosis in both pulmonary arteries and veins. The right ventricle, already having become hypertrophic, eventually fails; the pulmonary blood flow decreases, with associated reduction of the systemic blood flow. If the reduction of the cardiac output is critical, end organ failure with hepatic and/or renal insufficiency, cardiogenic shock and metabolic acidosis can occur. The right ventricular failure, with the frequently associated tricuspid valve regurgitation, is accompanied by systemic venous congestion with subsequent hepatomegaly, ascites and peripheral edema.

The presence of an associated atrial septal defect may temporary decompress the left

atrium, thereby reducing or masking the severity of the mitral stenosis. Associated malformations with post-mitral left-to-right shunt, like ventricular septal defect or patent ductus arteriosus, by increasing the pulmonary blood flow and therefore the pulmonary venous return can exacerbate the manifestations of obstruction to the mitral inflow.

Supravalvular mitral stenosis: the pathophysiologic pattern is exactly the same as for valvular stenosis, with variable degrees of obstruction to the mitral valve inflow. Very rarely the presence of an incomplete and eccentric supramitral ring allows unobstructed flow through the mitral valve. A supravalvular mitral ring can be very difficult to detect in the presence of associated malformation with reduced pulmonary blood flow, like tetralogy of Fallot.

Diagnosis

Clinical pattern:

- neonates with severe mitral stenosis present with respiratory distress from pulmonary edema shortly after birth if an unrestrictive interatrial communication does not exist; the presence of an atrial septal defect decompresses the left atrium with the subsequent clinical pattern of increased pulmonary blood flow and reduced systemic cardiac output;
- patients with a mild to moderate degree of mitral stenosis present after the neonatal period with signs of low cardiac output, recurrent pulmonary infections, poor feeding, failure to thrive, limited exercise tolerance, diaphoresis, tachypnea;
- older children may present with pulmonary congestion, progressive dyspnea, nocturnal cough, signs of right heart failure; they may develop atrial fibrillation, thromboembolic events (with the possibility of a stroke), infective endocarditis, hemoptysis (caused by rupture of dilated bronchial veins), chest pain, dysphagia (due to esophageal compression by the dilated left atrium);
- cardiac auscultation: loud first sound (abrupt closure of the mitral valve), accentuated intensity of second sound (pulmonary hypertension), long low-frequency diastolic murmur, best heard at the apex; opening snap of the mitral valve is not heard in supravalvular ring; in severe mitral valve stenosis there are diminished peripheral pulses and gallop murmur.

Electrocardiogram: left atrial enlargement and right ventricular enlargement and/or hypertrophy in proportion with the severity of the obstruction.

Chest X-ray: left atrial dilatation, straightening of the left cardiac border, widening of the tracheal carina, elevation of the left bronchus, prominent upper pulmonary veins, increased interstitial markings, Kerley lines, pulmonary artery trunk and branches dilatation, right ventricular dilatation (Fig. 2.9.1); barium-swallow study shows in the lateral projection a rounded indentation on the anterior wall of the esophagus.

Echocardiogram: is the most important diagnostic tool to evaluate patients with mitral stenosis; the entire mitral valve apparatus is

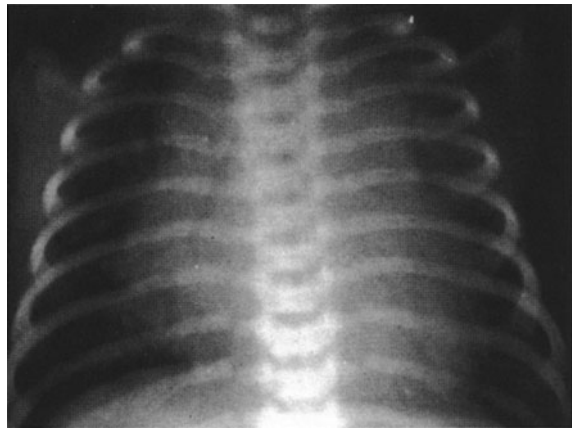

Fig. 2.9.1. Mitral valve disease: radiography. Anteroposterior view in an infant with severe mitral valve stenosis, showing left atrial dilatation, straightening of the left cardiac border, elevation of the left bronchus, prominent upper pulmonary veins, increased interstitial markings, Kerley lines, pulmonary artery trunk and branches dilatation, right ventricular dilatation

investigated, including morphology and function, with parasternal subcostal views (Fig. 2.9.2 and 2.9.3); the Doppler technique allows calculation of the transvalvular pressure gradient by measuring the mean velocity of diastolic flow through the valve.

Cardiac catheterization: not needed to diagnose mitral valve stenosis or supravalvular mitral ring, well defined and quantified by echocardiography; it allows direct measurement of the intracardiac pressures, and it can be useful in the presence of associated malformations not well demonstrated by echocardiography, particularly with regard to the presence of obstructions at the level of the aortic arch and isthmus.

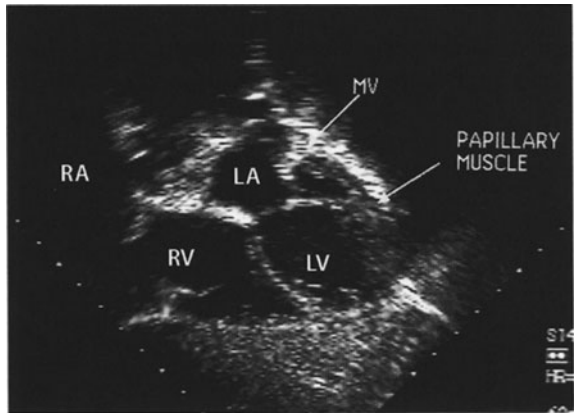

Fig. 2.9.2. Mitral valve disease: echocardiography. Subcostal 4-chamber view showing a parachute mitral valve (*LA* left atrium, *LV* left ventricle, *MV* mitral valve, *RA* right atrium, *RV* right ventricle) (photograph courtesy of Dr. Nicole Sekarski)

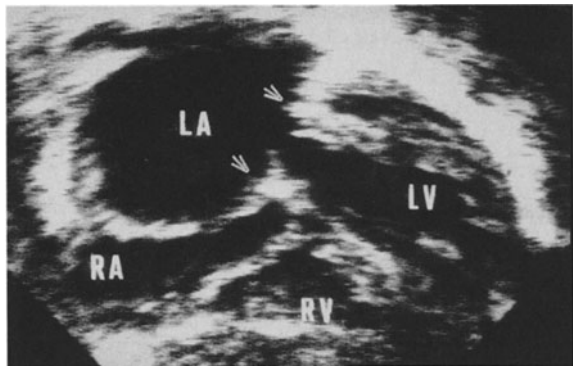

Fig. 2.9.3. Mitral valve disease: echocardiography. Subcostal 4-chamber view showing an arcade mitral valve (indicated by the two white arrows) (*LA* left atrium, *LV* left ventricle, *RA* right atrium, *RV* right ventricle) (reproduced with permission from Marino B, Thiene G (1990) Atlante di anatomia ecocardiografica delle cardiopatie congenite, USES, Firenze)

Mitral valve regurgitation

Morphology

As for mitral stenosis, congenital mitral regurgitation can present in several forms, due to the presence of malformations of one or more of the four major components of the mitral valve: the mitral annulus, the mitral leaflets, the chordae tendineae and the papillary muscles. All four anatomic components function to maintain competent mitral valve closure during systole, with decreased circumference of the valve annulus by 20 to 30% throughout systole, and contraction of the papillary muscle to maintain the length of the chordae under the pressure that develops during systole. In the presence of any developmental abnormality of one or more of the four components, mitral valve regurgitation results.

Mitral valve regurgitation, or with prevalent regurgitation (25–30% of patients), as a congenital malformation, is most frequently due to one of the following reasons: annular dilatation secondary to anterior or posterior leaflet prolapse, annular dilatation secondary to posterior leaflet hypoplasia with chordal shortening, isolated mitral cleft (not associated with atrioventricular septal defect) or fenestrations of the anterior leaflet, valve prolapse due to elongated chordae, hammock valve or anomalous mitral arcade. Congenital isolated annular dilatation is extremely rare.

Mitral valve with stenosis and regurgitation, or with combined lesions (15% of patients) may result from a variety of combinations of malformations leading to mitral valve stenosis and/or regurgitation.

Associated anomalies

Mitral valve regurgitation, frequently associated with atrial or ventricular septal defect, can rarely be associated with subaortic obstruction, aortic coarctation, patent ductus arteriosus, double outlet right ventricle (particularly with subpulmonary ventricular septal defect), transposition of the great arteries, anomalous origin of the left coronary artery, pulmonary valve stenosis. Mitral valve malformations are also present in Marfan syndrome, Ehlers-Danlos syndrome, and in endocardial fibroelastosis.

Pathophysiology

The presence of mitral regurgitation allows for return of blood flow from the left ventricle to the left atrium in systole. As the volume of the regurgitation increases with time, the left ventricle also increases in size. The progressive dilatation of the left ventricle due to the volume overload, further increasing the mitral valve regurgitation because of the associated dilatation of the valve annulus, eventually leads to impaired ventricular contraction, increased afterload, reduced cardiac output, and, finally, left heart failure.

The pathophysiologic pattern is characterized by the simultaneous presence in systole of antegrade blood flow from the left ventricle to the aorta and another volume, the regurgitant fraction, retrograde blood flow from the left ventricle to the left atrium. This causes a proportionate increase of the left ventricular ejection volume. The regurgitant fraction returns to the left ventricle in diastole producing a ventricular volume overload. The compensation by the left ventricle occurs thanks to the Frank-Starling mechanism, resulting in a greater ventricular stroke volume.

The natural history and time course of mitral regurgitation is variable, but mitral regurgitation can develop in three distinct stages, each one with relevant clinical significance: acute, chronic compensated and chronic decompensated. The stages depend on the rapidity of the beginning, amount of regurgitant volume and left atrial compliance.

Acute mitral regurgitation stage: acute mitral regurgitation causes sudden volume overload of the left atrium and left ventricle. Initially the nondilated left atrium limits the regurgitant volume at the expense of an increase in both left atrial and left ventricular enddiastolic pressures. Although total ventricular stroke volume increases compared to normal, total forward stroke volume usually decreases, thereby reducing cardiac output. In the acute situation, rapidly increasing left atrial pressure results in elevated pulmonary venous pressure causing pulmonary congestion and, eventually, pulmonary edema.

Chronic compensated stage: in this stage the left ventricle compensates by allowing greater diastolic filling and developing left ventricular enlargement to augment forward stroke volume. More importantly, the left atrium dilates in response to the increased volume overload. Compensation for the increased volume can occur without resulting in increased pressure in the pulmonary circulation and the right heart. Left atrial compliance decreases the left ventricular afterload, while left ventricular dilation and hypertrophy increases the contractility. These important changes keep the overall afterload on the left heart normal or unchanged. Although the regurgitant fraction may be high, the larger stroke volume compensates, maintaining a nearly normal forward cardiac output.

Chronic decompensated stage: this stage occurs when the left ventricle becomes unable to sustain adequate forward cardiac output. As the left ventricular contractility begins to decrease, the endsystolic volume gradually increases, thereby increasing the left ventricular enddiastolic pressure. The resulting increased pressure in the left atrium creates increased afterload, which further impairs the left ventricular ejection, thereby creating a vicious cycle. While the enddiastolic and endsystolic volumes increase, pulmonary congestion

eventually results if the cause of the mitral regurgitation is not removed. Although the forward left ventricular ejection fraction is reduced compared to the compensated phase, the overall ejection fraction could remain normal because of a large regurgitant flow. As the degree of mitral regurgitation worsens, the total ejection fraction falls, indicating increasing ventricular dysfunction. Pulmonary hypertension may develop under long-standing increased pulmonary venous pressure, and, ultimately, it can lead to right heart failure.

Diagnosis

Clinical pattern:
- *acute severe mitral regurgitation:* children may present in heart failure or cardiogenic shock;
- *chronic mitral regurgitation:* depending upon the degree of regurgitation, children may be asymptomatic and remain so for many years, or with only mild fatigue; with moderate to severe regurgitation, tachypnea and dyspnea may occur, limited growth and failure to thrive; hemoptysis can develop;
- on physical examination cardiac impulse may be displaced to the left and peripheral pulses may have small volume and sharp upstroke;
- on auscultation usually the first sound is slightly diminished, while the second sound is split; there is a olosystolic murmur, loudest over the apex with radiation to the left axilla; with pulmonary hypertension the second sound becomes louder.

Electrocardiogram: left atrial and left ventricular enlargement; in the presence of pulmonary hypertension: right ventricular hypertrophy; rhythm disturbances, like atrial fibrillation, are extremely rare in children.

Chest X-ray: cardiomegaly associated with the degree of valve stenosis or regurgitation;
- *chronic mitral valve regurgitation:* left atrial and left ventricular enlargement, pulmonary congestion;

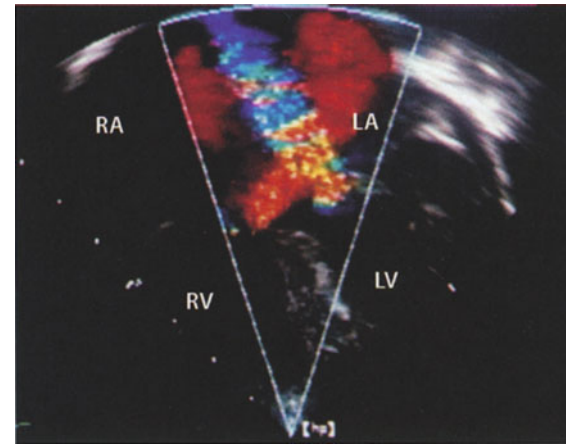

Fig. 2.9.4. Mitral valve disease: echocardiography. A 4-chamber view with color Doppler showing the mitral valve regurgitation extended into the entire left atrium (*LA* left atrium, *LV* left ventricle, *RA* right atrium, *RV* right ventricle) (photograph courtesy of Dr. Michael Rigby)

- *acute mitral valve regurgitation:* pulmonary venous vasculature markings, pulmonary edema.

Echocardiogram: is the best diagnostic technique to evaluate mitral valve morphology and function; the Doppler color flow in precordial short-and long-axis views demonstrates duration and direction of the regurgitant flow, with high-velocity systolic signal across the valve; grading of the regurgitation (Fig. 2.9.4) as mild, moderate or severe is based on the size and extent of the regurgitation into the left atrium; *mild*: proximal third of the left atrium near the mitral valve annulus; *moderate*: mid left atrial cavity; *severe*: posterior third of the left atrium, including the pulmonary veins.

Cardiac catheterization: it allows direct measurement of the intracardiac pressures; indicated in the presence of discordance between the clinical and echocardiographic data.

Double orifice mitral valve

Morphology

Rare malformation where the mitral valve presents with a single fibrous annulus with two separate orifices, each supported by its own tensor apparatus, opening into the left ventricle. The subvalvular structures invariably show varying degrees of abnormality, particularly at the level of the tensor apparatus. There are three major types of double orifice mitral valve:

Eccentric or hole type: this is the most frequent variety (about 80% of the cases) and it presents with a small accessory orifice located at either the anterolateral or posteromedial commissura. Generally these are associated with other malformations of the valve apparatus, like cleft leaflets, accessory or fused papillary muscles, and crossing chordae tendineae.

Central or bridge type: in about 15% of the patients a central bridge of fibrous or abnormal leaflet tissue connects the two leaflets of the mitral valve, dividing the orifice into medial and lateral portions; these two openings may be equal of different; the papillary muscles are generally normal with chordae surrounding each orifice inserting into only one papillary muscle.

Duplicate mitral valve: in this extremely rare type, there are two mitral valve annuli and valves, each with its own set of leaflets, commissurae, chordae and papillary muscles.

Associated anomalies

The most frequent associated cardiac lesion is the atrioventricular septal defect, particularly when the accessory orifice is located at the posteromedial commissural. Other associated cardiac malformations include ventricular septal defect, subaortic obstruction, patent ductus arteriosus, aortic coarctation and aortic arch interruption. Rare associated lesions are Ebstein's anomaly, parachute mitral valve, tetralogy of Fallot, hypoplastic left heart syndrome, bicuspid aortic valve, pulmonary valve stenosis, truncus arteriosus.

Pathophysiology

While it is possible that a double orifice mitral valve allows normal hemodynamic flow between the left atrium and the left ventricle (in less than 50% of the patients), most fre-

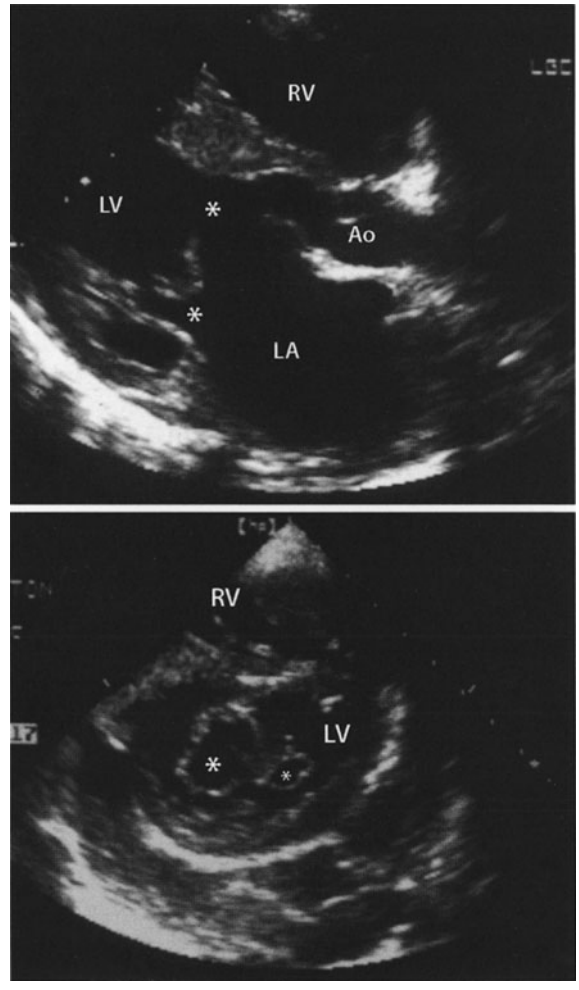

Fig. 2.9.5. Mitral valve disease: echocardiography. **a** Parasternal long-axis view showing a double orifice (large and small white asterisks) mitral valve, **b** parasternal short-axis view of the same patient showing a double orifice (large and small white asterisks) mitral valve (photographs courtesy of Dr. Michael Rigby)

quently it may obstruct the mitral valve inflow and/or determine mitral valve regurgitation. The combined area of the double orifice mitral valve may be significantly less than in a normal mitral valve. The reduction of the effective valve area can be determined by structural abnormalities including large bridging tissue, bulky abnormal leaflets, fused chordae and abnormal papillary muscles. With the association of complete atrioventricular septal defect, the combined area can reach 85–90% of the normal area.

Diagnosis

Clinical pattern, electrocardiogram and chest X-ray are very similar to the other cases with either mitral stenosis or regurgitation.

Echocardiogram: the two orifices of double orifice mitral valve are best visualized in short-axis cross sectional view (Fig. 2.9.5), scanning from the left ventricular apex to the base; apical and subcostal 4-chamber views are also useful to visualize the subvalvular apparatus; Doppler color flow shows the flow pattern through the mitral valve.

Mitral valve disease

Indications for surgical treatment

Indication for surgery is straightforward in all symptomatic children. In asymptomatic patients the decision must take into consideration the mismatch between the morphology of the valve and the surgical experience with reparative valve procedures. Congenital mitral valve defects remain a surgical challenge, particularly in mitral stenosis, and when associated with other heart defects, because of a wide variety of apparatus anomalies and the young age of the patients, and because there is no ideal substitute for the mitral valve, and repair is not always feasible.

Reconstructive surgery is always the primary goal, and valve replacement as first stage approach should be considered as a salvage procedure, particularly in the youngest patients.

Mitral stenosis: mitral valve commissurotomy, division of fused chordae tendineae and papillary muscles can be taken into consideration.

Mitral regurgitation: partial plication annuloplasty is the main technique for congenital mitral regurgitation because this technique allows the mitral annulus to grow, in contrast to ring annuloplasty; the only limit of this technique is the absence of chordae, where artificial chordae replacement provides better long-term results. Partial annuloplasty, repair of the cleft of the anterior leaflet, chordal shortening, chordal transfer, artificial chordae, resection or reconstruction of the anterior or posterior leaflet, modified de Vega are all techniques available to the armamentarium of cardiac surgeons in this situation. Accessory mitral valve tissue causing left ventricular outflow tract obstruction may require surgical removal only in the presence of severe obstruction.

Surgical treatment (on cardiopulmonary bypass)

Both the procedures of mitral valve repair and replacement are performed on cardiopulmonary bypass, with approach to the left atrium either directly with an incision parallel and posterior to the interatrial groove, or on the dome of the left atrium, or through an incision of the interatrial septum, after right atriotomy. In small infants a combination of the above incisions may be necessary to obtain adequate surgical exposure.

Since the pathology of congenital mitral malformations is extremely variable, it is impossible to provide details of the various surgical techniques, particularly because they all depend upon the mismatch between

the anatomy of the valve and the experience of the individual surgeon. Because of the negative aspects of mitral valve replacement (distortion of the left ventricular geometry, impairment of the left ventricular function, need for anticoagulation), particularly in young patients (higher mortality, outgrowth of the valve), and because of the unavailability of small size prosthetic valves (the smallest commercially available mechanical valve is a 15 mm diameter prosthesis), aggressive reconstructive procedures are always taken into consideration to avoid valve replacement, particularly in the first few years of life.

Mitral valve repair:

- *Mitral stenosis:* the fused mitral valve commissurae are directly incised with full mobilization of the leaflets, and the thickened leaflets are shaved; fused chordae tendineae are incised and split as well as the fused papillary muscles to relieve subvalvular stenosis; the supravalvular ring is completely resected. Resection of accessory mitral valve tissue causing left ventricular outflow tract obstruction can be performed through a transatrial or transaortic approach, or from the combined approach.
- *Mitral regurgitation:* Partial annuloplasty, repair of the cleft of the anterior leaflet, chordal shortening, chordal transfer, artificial chordae (5–0 expanded PTFE sutures) to replace absent, fused or elongated chordae, resection or reconstruction of the anterior or posterior leaflet with pericardial patch leaflet extension, modified de Vega are all techniques available, mostly similar to the techniques used in adult patients.

  After mitral valve repair, the obtained diameter is measured with Hegar dilators and compared with the normal value for age and body weight; then a static test to rule out residual or induced regurgitation is performed with injection of saline solution under pressure into the left ventricular chamber; the most reliable evaluation is obtained with intraoperative transesophageal echocardiography after weaning from cardiopulmonary bypass.

Mitral valve replacement: the surgical technique has to take in consideration the mismatch between the size of the patient and the size of the available prosthetic valve. The approach is the same as for mitral valve repair, but in the smallest patients it is necessary to implant the prosthetic valve in the supraannular position in the left atrium, in strict proximity of the orifices of the pulmonary veins.

When a mitral valve replacement is unavoidable, the choice among the available options is relatively limited, particularly in smaller children:

- *Biological valves* are not an ideal substitute, because of early calcification, due to the accelerated calcium metabolism in children, and biological tissue degeneration. The experience with homografts for mitral valve replacement in children is still too limited and with a reduced applicability.
- *Mechanical valves* required life-long anticoagulation and present the problem of outgrowth; bileaflet mechanical prostheses are the valve of choice in infants and small children.
- *Homografts* can be used, but the experience with homografts for mitral valve replacement in children is still too limited and with a reduced applicability, and furthermore with poor medium and long-term results, because of thickening, shrinking and calcification of the homograft implanted in the systemic circulation.

Alternative surgical techniques have been utilized:

- *Pulmonary autograft* (Kabbani-Ross mitral procedure). A pulmonary autograft, like for the Ross procedure (see chapter "Left ventricular outflow tract obstruction"), is used to replace the malformed mitral valve in neonates and infants unsuitable

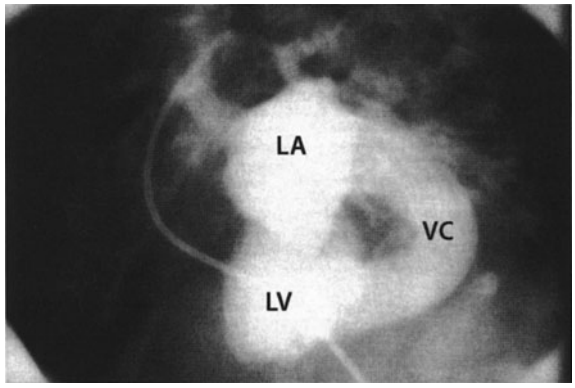

Fig. 2.9.6. Mitral valve disease: surgery. Postoperative angiography in left anterior oblique view showing a valved conduit implanted from the left atrium to the left ventricle to bypass a hypoplastic mitral valve (*LA* left atrium, *LV* left ventricle, *VC* valved conduit) (reproduced with permission from Corno AF, Giannico S, Leibovich S, Mazzera E, Marcelletti C (1986) The hypoplastic mitral valve. When should a left atrial left ventricular extracardiac valved conduit be used? J Thorac Cardiovasc Surg 91:848–851)

for a conventional approach; of course a biological valved conduit is needed to replace the explanted native pulmonary valve.

- *Valved conduit bypass of the mitral valve.* In the presence of severely hypoplastic mitral valve annulus, where conventional reconstructive procedures are inadequate and valve replacement is impossible because of the small size of the mitral valve annulus, bypass of the mitral valve is possible by means of a valved conduit implanted between the left atrium and the left ventricle; the proximal anastomosis is performed on the incised left auricular appendage and the distal anastomosis on an apical left ventriculotomy parallel to the left anterior descending coronary artery (Figs. 2.9.6 and 2.9.7).

Potential complications

Mitral valve repair: residual or recurrent mitral valve stenosis or regurgitation, requiring reoperation. Particularly less satisfactory are the results of repair of the hammock mitral valve.

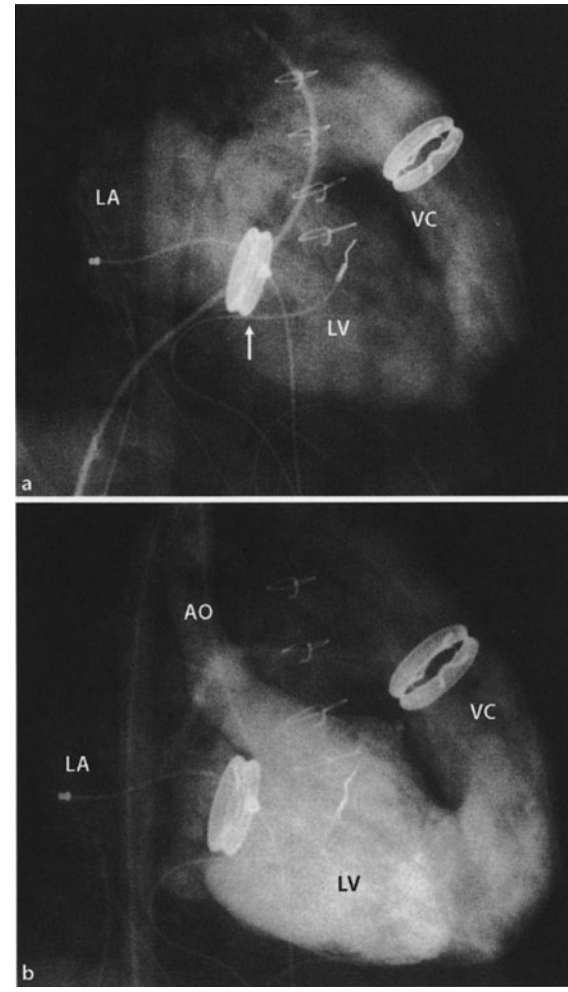

Fig. 2.9.7. Mitral valve disease: surgery. Postoperative angiography in left anterior oblique view with pulmonary artery injection **a** showing a valved conduit implanted from the left atrium to the left ventricle to bypass an outgrown prosthetic mitral valve (white arrow) and **b** showing the left ventricular filling through the valved conduit implanted from the left atrium to the left ventricle and the opacification of the aorta

Mitral valve replacement: in younger children (less than 3–5 years of age) it is still accompanied by elevated mortality and morbidity, substantially higher than in older children and young adults. Potential complications include complete atrioventricular block, paravalvular leak, thromboembolism, bleeding, arrhythmias, prosthetic valve endocarditis, structural and nonstructural (valve entrapment by pannus formation) prosthetic valve dysfunction, patient-valve mismatch, patient outgrowth of the artificial prosthesis.

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Chapter 2.10 Aneurysm of Valsalva

Incidence

Aneurysms of sinus of Valsalva are very rare congenital malformations, with an incidence between 0.1 and 3.5% of all congenital heart defects. The incidence is more elevated among the Asian population, with an incidence up to five times higher than in Western population, with a 2:1 to 4:1 male to female ratio, because of the higher incidence of infundibular septal defects; in fact this type of defect may contribute to the instability of the aortic sinuses, particularly of the right sinus.

Morphology

The sinuses of Valsalva are located in the most proximal part of aorta, just above the cusps of the aortic valve, and end at the level of the sino-tubular junction, where the tubular portion of the aorta begins, each sinus corresponding to an individual cusp of the aortic valve.

The aneurysm of sinus of Valsalva is a congenital disorder consisting of a weak point in the aortic wall resulting from a localized interruption of the media within one of the sinuses, adjacent to the hinge line of the cusp of the aortic valve; the consequence is a thin-walled saccular or tubular outpouching, like a windsock. It occurs more frequently in the right coronary sinus (80–90% versus 65–70%, respectively in the Asian and Western population) than in the noncoronary sinus (10–20% versus 20–35%, respectively in the Asian and Western population). Most of the reported cases (65%) came to observation because of the rupture of the aneurysm, in comparison with a smaller number of cases (35%) diagnosed without rupture. Due to the central position of the aortic root, the congenital aneurysm of sinus of Valsalva can rupture into any of the cardiac chambers: the most frequent are right ventricle (94% versus 57%, respectively in the Asian and Western population of the cases) and right atrium (between 5% and 30%); rarely (equal or less than 1%) rupture has been reported in left atrium, left ventricle or pulmonary artery.

Associated anomalies

Ventricular septal defect, particularly the infundibular type, is frequently (50–60% versus 35–40% of the cases, respectively in the Asian and Western population) associated particularly with aneurysm of the right coronary sinus, less frequently aortic valve regurgitation (20–30% of the cases), left ventricular outflow tract obstruction with subaortic membrane or bicuspid aortic valve (10%), pulmonary stenosis (5%), atrial septal defect (2–5%), aortic coarctation (1–2%); occasional association has been reported with tetralogy of Fallot, anomalous origin of a coronary artery, patent ductus arteriosus.

Various generalized disorders may be associated with dilatation and/or distortion of the aortic root, including Marfan syndrome, Ehlers-Danlos syndrome, Turner syndrome, Williams syndrome and osteogenesis imperfecta.

A. F. Corno, *Congenital Heart Defects*

Pathophysiology

Aneurysmal dilatation of the sinuses of Valsalva occurs when the aortic media is defective, allowing separation of the media from the fibrous aortic annulus. Under the strain of aortic pressure, the involved sinus gradually weakens and dilates like a windsock, causing the formation of the aneurysm. Lack of supporting tissue, like in the presence of infundibular septal defect, may contribute to instability and progressive distortion of the aortic sinus, often with associated aortic valve regurgitation. Distortion and prolapse of the involved sinus and of the aortic valve leaflet can lead to progressive aortic valve regurgitation.

Generally asymptomatic, the aneurysm of sinus of Valsalva becomes evident when rupture occurs. At this point an acute left-to-right shunt from the aorta to one of the right cavities leads to the development of ventricular volume overload with simultaneous acute aortic valve regurgitation. Rupture may occur into any cardiac cavity, although most frequently occurs into a low-pressure chamber, the right ventricle; rupture into the right atrium is the second most common, followed by rupure into the left-sided cavities; extremely rarely rupture occurs into the pericardium. The pathophysiologic pattern depends upon size and location of the shunt resulting from the rupture of the aneurysm. Right ventricular outflow tract obstruction, coronary artery compression, left atrial roof compression with subsequent mitral valve regurgitation, ventricular tachycardia and complete atrioventricular block have been reported as consequences of the presence of an aneurysm of sinus of Valsalva. Endocarditis or thromboembolism can also be the initial manifestation of sinus of Valsalva.

Diagnosis

Clinical pattern: important aneurysmal dilatation is rarely seen at birth; generally asymptomatic until rupture occurs, with acute onset of overwhelming congestive heart failure, cardiac tamponade, arrhythmias, myocardial ischemia, depending upon the size and location of the aneurysm and the subsequence flow disturbance; 15–20% of patients with ruptured aneurysm of Valsalva remain asymptomatic; sudden death can follow rupture of an aneurysm of Valsalva, because of any of the above problems; young adults with unruptured aneurysm may present with effort dyspnea, chest pain, palpitations, signs of advancing heart failure; clinical signs may be absent or due to the left-to-right shunt with aortic valve regurgitation: widened pulse pressure, bounding peripheral pulses, precordial thrill, and loud continuous murmur, accentuated in diastole, along the left sternal border.

Electrocardiogram: left or right ventricular or biventricular enlargement, right bundle branch block are frequent; myocardial ischemia with depression of the ST-T segment and conduction disturbances are less frequent.

Chest X-ray: cardiomegaly, with right heart enlargement and increased lung vascularity in the presence of rupture into the right ventricle or right atrium, and with enlargement of the aortic root in the case of rupture into the left ventricle.

Echocardiogram: is the most reliable method to confirm the diagnosis, showing the morphology of the proximal aorta and of the aortic sinuses (Fig. 2.10.1); Doppler color flow can indicate and quantitate the shunt due to the rupture of the aneurysm, and also the degree of aortic valve regurgitation; transesophageal echocardiography provides better anatomical and functional characterization of the defect (Fig. 2.10.2).

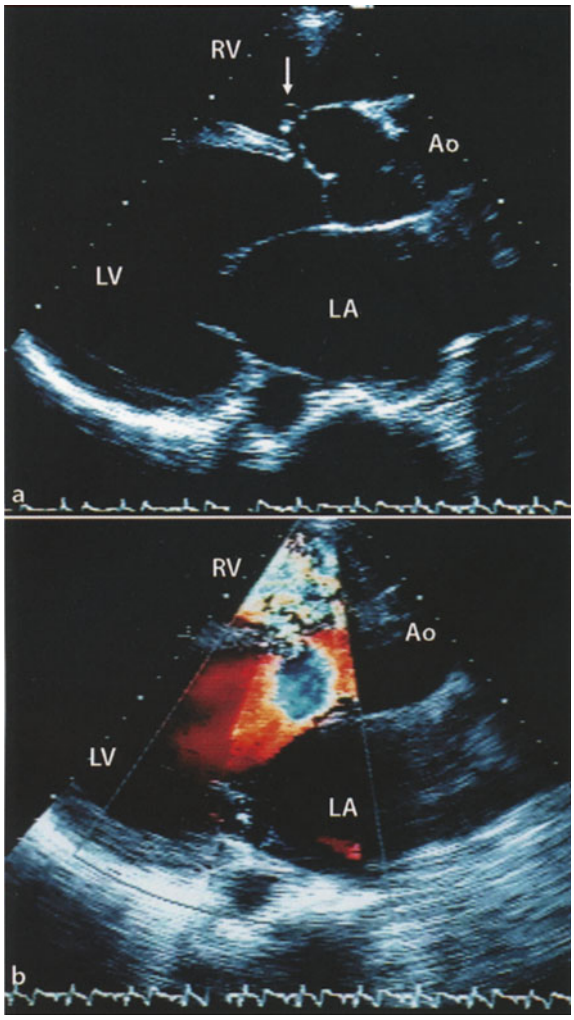

Fig. 2.10.1. Aneurysm of Valsalva: **a** preoperative transthoracic echocardiography, showing the ruptured aneurysm of the right sinus of Valsalva (arrow) protruding into the right ventricle through a large ventricular septal defect, **b** preoperative transthoracic Doppler echocardiography of the same patient, showing the severe regurgitation into the left ventricle from the ruptured aneurysm of the right sinus of Valsalva and the left-to-right shunt through the ventricular septal defect (*Ao* aorta, *LA* left atrium, *LV* left ventricle, *RV* right ventricle)

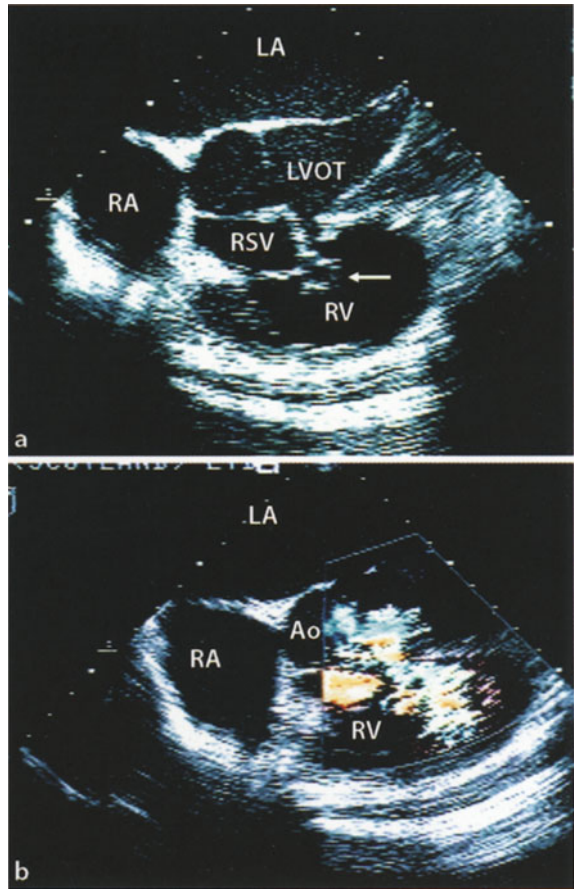

Fig. 2.10.2. Aneurysm of Valsalva: **a** intraoperative transesophageal echocardiography of the same patient as in Fig. 2.10.1, showing the ruptured aneurysm of the right sinus of Valsalva (arrow) protruding into the right ventricle through a large ventricular septal defect (*LA* left atrium, *LVOT* left ventricular outflow tract, *RA* right atrium, *RSV* right sinus of Valsalva, *RV* right ventricle), **b** intraoperative transesophageal Doppler echocardiography of the same patient, showing the left-to-right shunt aorta-to-right ventricle from the ruptured aneurysm of the right sinus of Valsalva (*Ao* aorta)

Cardiac catheterization: angiography can be useful to detect coronary artery anomalies or compression.

Indications for surgical treatment

Because of the sudden hemodynamic deterioration in case of rupture, with most patients coming to death within a short period after rupture, early diagnosis is mandatory and prompt surgical treatment is often life-saving.

In patients with unruptured aneurysm, indication for surgery depends upon the associated lesions and the degree of aortic valve regurgitation, and of course upon the subsequent hemodynamic pattern and the degree and rapidity of progressive enlargement of the aneurysm itself. Unfortunately in most patients the time-related probability of aneurysmal rupture is impossible to determine based on the clinical history. Furthermore

the natural history or unruptured aneurysm of sinus of Valsalva is unknown, because most of these lesions are asymptomatic and remain undetected.

Despite recent reports of closure of ruptured aneurysms of Valsalva with a procedure of interventional cardiology, surgical repair remains the gold standard to obtain complete closure and to avoid recurrence.

Surgical treatment (on cardiopulmonary bypass)

Surgery of ruptured aneurysms is performed on cardiopulmonary bypass, with particular attention to the administration of the cardioplegic solution, preferable through retrograde infusion into the coronary sinus. The surgical repair is accomplished with direct or patch closure of the rupture, either from an single approach through an aortotomy (Fig. 2.10.3) or through an incision in the cardiac chamber of the exit of the aneurysm, or from double approach involving both the involved cardiac chamber and the aortic root.

The technique with patch closure is the most reliable because it is accompanied by the lowest rate of recurrencies. Of course perfect exposure is needed to visualize the defect as well as the aortic valve leaflets and to identify the orifice of the coronary arteries.

While associated ventricular septal defect requires patch closure, even for relatively small defects, to avoid recurrencies (see chapter "Ventricular septal defect"), the repair of the aortic valve is the first choice, particularly in the pediatric age, with direct or patch resuspension of the aortic valve leaflets; again, patch technique provides better long-term results in terms of reducing the need for aortic valve reoperation or replacement. The surgical exposure of the ventricular septal defect can be obtained either through the same aortotomy used to expose the aneurysm, or through an incision in the main pulmonary artery, since in the vast majority of the patients it is an infundibular type of ventricular septal defect; very rarely a right ventriculotomy is required.

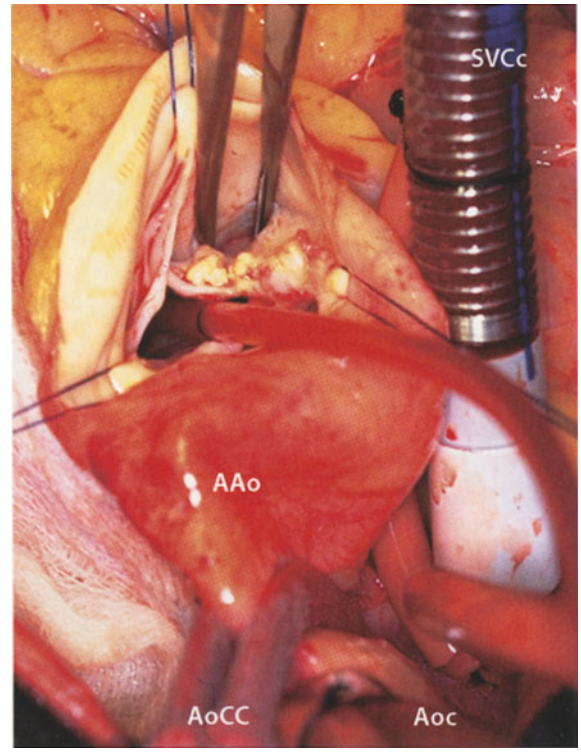

Fig. 2.10.3. Aneurysm of Valsalva: surgery. Intraoperative photograph showing a rupture of the non-coronary sinus of Valsalva, indicated by the open forceps; the aortic valve is exposed with three stay sutures at the level of the three commissurae, and a pump sucker is introduced into the left ventricle through the open aortic valve (*AAo* ascending aorta, *AoCC* aortic cross clamp, *Aoc* aortic cannula, *SVCc* superior vena cava cannula)

The surgical repair of unruptured aneurysms is generally performed through only an aortotomy.

Associated aortic valve replacement can be required in the presence of severe distortion of the aortic root or involvement of the aortic valve, that is not suitable to a valve-sparing procedure, either at the moment of the aneurysm repair or later during the follow-up, due to progression of the aortic valve regurgitation.

Intraoperative transesophageal echocardiography not only allows adequate preoperative evaluation of the defect, but it is essential to control the quality of the surgical repair, in particular regarding the presence of

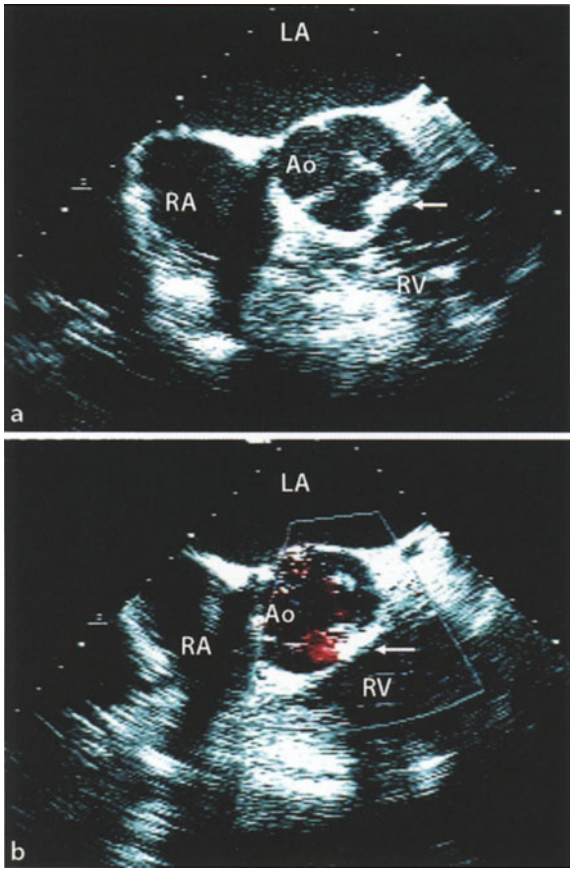

Fig. 2.10.4. Aneurysm of Valsalva: surgery. **a** Postoperative transesophageal echocardiography of the same patient as in Figs. 2.10.1 and 2.10.2, showing the patch closure (arrow) of the ruptured aneurysm of the right sinus of Valsalva, **b** postoperative transesophageal Doppler echocardiography of the same patient, showing the abolished left-to-right shunt due to the patch closure (arrow) of the ruptured aneurysm of the right sinus of Valsalva (*Ao* aorta, *LA* left atrium, *RA* right atrium, *RV* right ventricle)

residual fistulous communication, residual ventricular septal defect or aortic valve regurgitation (Fig. 2.10.4).

Potential complications

Residual or recurrent shunt from the aorta to the affected cardiac chamber, residual or progressive aortic valve regurgitation, residual or recurrent ventricular septal defect, complete atrio-ventricular block, arrhythmias, myocardial infarction, infective endocarditis, thromboembolism.

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Chapter 2.11 Double outlet right ventricle

Incidence

Double outlet right ventricle is the 12th most common congenital heart defect (1.5–2.0% of all congenital heart defects). Occurs in 0.03–0.1/1,000 live births. No sex prevalence is reported.

Morphology

Double outlet right ventricle defines a heterogeneous group of cardiac malformations unified by an abnormal ventriculoarterial connection. The general agreement is about the definition of double outlet right ventricle when both great arteries, or one of the two great arteries and more than half of the other great artery originate from the right ventricle.

According to Yves Lecompte, the classification and terminology of this complex group of patients with anomalous ventriculoarterial connection is less important than the precise preoperative definition of the anatomic criteria useful to determine the best surgical approach. Nevertheless the categorization of patients with double outlet right ventricle is necessary to compare the results of different surgical treatments.

A morphological feature characteristic of the double outlet right ventricle is absence of the normal fibrous continuity between the mitral and semilunar valve (either the aortic valve in the presence of ventriculoarterial concordance, or the pulmonary valve in the presence of ventriculoarterial discordance), referred to as the presence of a, respectively, subaortic or subpulmonary conus.

The relationship between the great arteries may be in any of the various possibilities, but usually they are side by side and parallel. The aortic valve could be to the right or left of the pulmonary valve or in an anteroposterior relationship.

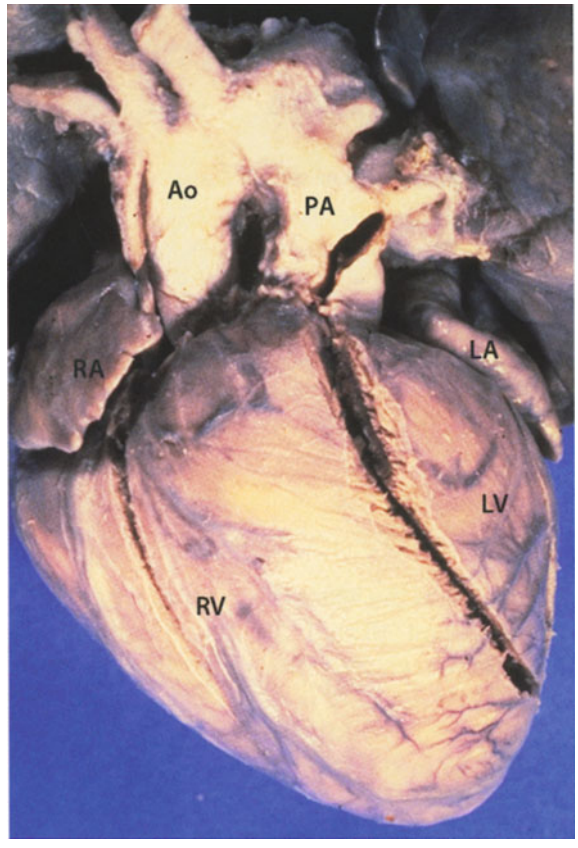

Fig. 2.11.1. Double outlet right ventricle: morphology. External appearance of a heart with double outlet right ventricle with ventriculoarterial concordance (*Ao* aorta, *LA* left atrium, *LV* left ventricle, *PA* pulmonary artery, *RA* right atrium, *RV* right ventricle) (photograph courtesy of Dr. Bruno Marino)

A. F. Corno, *Congenital Heart Defects*

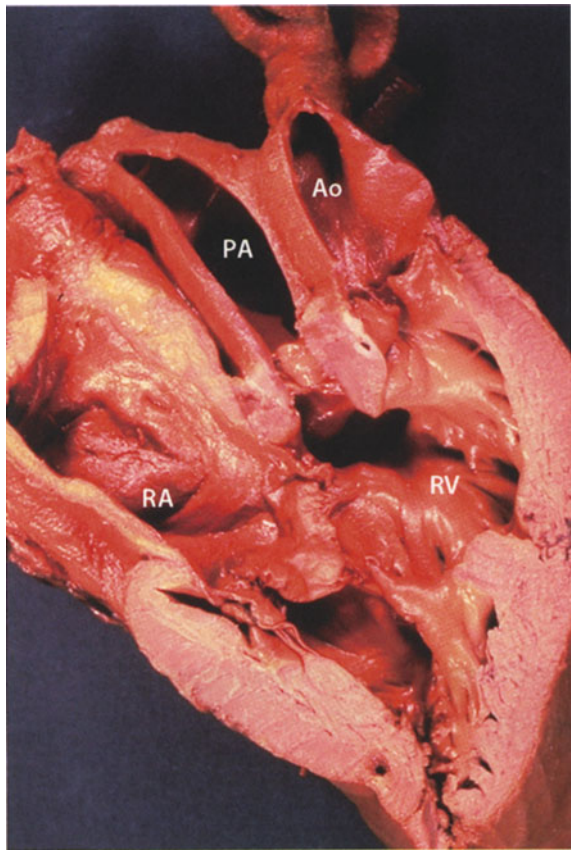

Fig. 2.11.2. Double outlet right ventricle: morphology. Subxiphoid right oblique view of a heart with double outlet right ventricle, ventriculoarterial discordance and subpulmonary ventricular septal defect (so-called Taussig-Bing malformation) (*Ao* aorta, *PA* pulmonary artery, *RA* right atrium, *RV* right atrium) (Reproduced with permission from Marino B, Thiene G (1990) Atlante di anatomia ecocardiografica delle cardiopatie congenite, USES, Firenze)

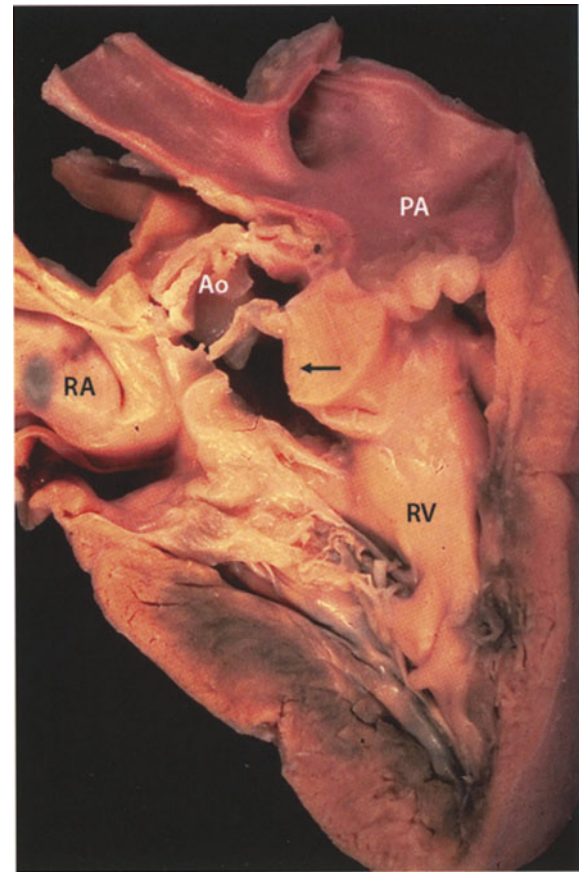

Fig. 2.11.3. Double outlet right ventricle: morphology. Right anterior oblique view of a heart with double outlet right ventricle with ventriculoarterial concordance and subaortic ventricular septal defect (black arrow) (*Ao* aorta, *PA* pulmonary artery, *RA* right atrium, *RV* right ventricle) (photograph courtesy of Dr. Bruno Marino)

Patients with double outlet right ventricle have been categorized according to:

The type of the ventriculoarterial connection:
- concordant (85%): the pulmonary artery originates entirely from the right ventricle (Fig. 2.11.1),
- discordant (15%): the aorta originates entirely from the right ventricle (Fig. 2.11.2).

The position of the associated ventricular septal defect:
- subaortic ventricular septal defect (Fig. 2.11.3) (50%),
- subpulmonary ventricular septal defect (Fig. 2.11.4) (30%),
- doubly-committed ventricular septal defect (immediately underneath the semilunar valves) (10%),
- noncommitted (remote) ventricular septal defect (far from both the semilunar valves) (10%).

The type of pulmonary blood flow
- restricted (= with pulmonary stenosis)
- unrestricted (= without pulmonary stenosis).

The most frequent combination is *double outlet right ventricle with concordant ventriculoarterial connection, subaortic ventricular septal defect and obstruction to the pulmo-*

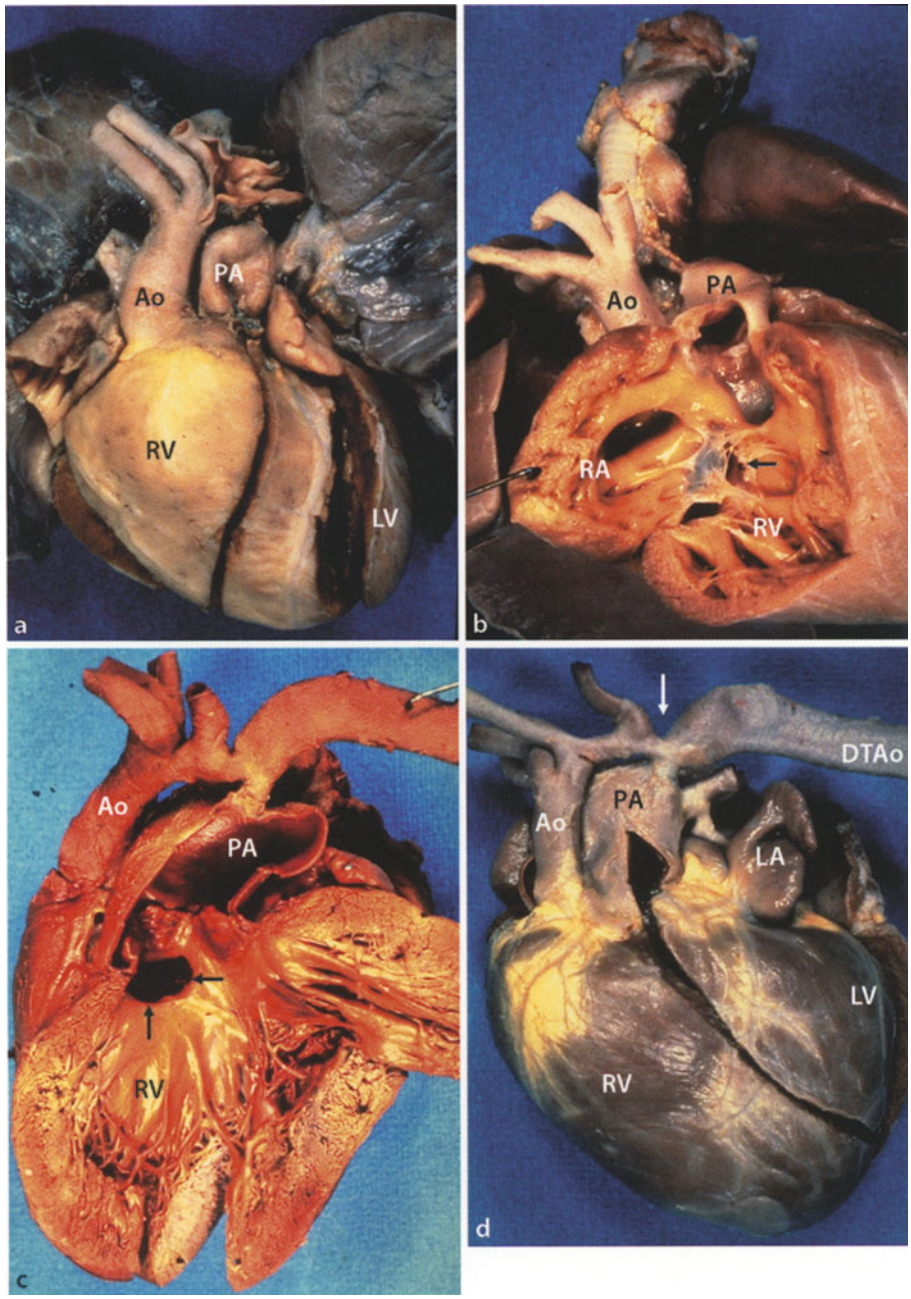

Fig. 2.11.4. Double outlet right ventricle: morphology. **a** External appearance in left oblique subxiphoid view of a heart with double outlet right ventricle, ventriculoarterial discordance and aortic coarctation (so-called Taussig-Bing malformation), **b** internal appearance in left oblique subxiphoid view of the same heart with double outlet right ventricle, ventriculoarterial discordance, subpulmonary ventricular septal defect (black arrow) and aortic coarctation (so-called Taussig-Bing malformation), **c** internal appearance in the lateral view of the same heart with double outlet right ventricle, ventriculoarterial discordance and aortic coarctation (so-called Taussig-Bing malformation), with the subpulmonary ventricular septal defect (black arrows), **d** external appearance of a heart with double outlet right ventricle, ventriculo-arterial discordance and aortic coarctation (white arrow) (so-called Taussig-Bing malformation) (*Ao* aorta, *DTAo* descending thoracic aorta, *LA* left atrium, *LV* left ventricle, *RA* right atrium, *RV* right ventricle, *PA* pulmonary artery) (photographs courtesy of Dr. Bruno Marino)

nary outflow tract (so-called tetralogy of Fallot-type). This type of double outlet right ventricle is morphologically and functionally very similar to the tetralogy of Fallot, in particular regarding the obstruction at the level of the right ventricular outflow tract. The main difference with tetralogy of Fallot is that it is possible (even if more rare, about 10% of the patients) to have a double outlet right ventricle of this type with restrictive ventricular septal defect, because of an anatomically small defect or because of the anomalous insertion of the tricuspid valve. The right ventricular outflow tract obstruction can be at the infundibular level (the most frequent), the valvular level (with or without annular hypoplasia), or both; there are also instances with isolated low level infundibular obstructions, producing a two-chambered right ventricle. Typical cardiac anomalies associated with this type of double outlet right ventricle are mitral stenosis and subaortic obstruction, as well as anomalous attachments (straddling) of the anterior and septal leaflets of the tricuspid valve.

The second most frequent type is *double outlet right ventricle with discordant ventriculoarterial connection and subpulmonary ventricular septal defect, without obstruction to the pulmonary outflow tract (so-called Taussig-Bing malformation).* In this type of double outlet right ventricle, the hemodynamic pattern is similar to the transposition of the great arteries with unrestrictive ventricular septal defect and malalignment of the infundibular septum, and the two malformations are quite frequently confused, since in double outlet right ventricle the pulmonary artery is overriding a large anterior ventricular septal defect, and the two great arteries are parallel to each other. Pulmonary stenosis is practically exceptional in this type. Typical cardiac anomalies associated with this type of double outlet right ventricle are straddling mitral valve, subaortic obstruction and aortic coarctation.

Another combination is *double outlet right ventricle with concordant ventriculoarterial connection and subaortic ventricular septal defect, without obstruction to the pulmonary outflow tract.* This type of double outlet right ventricle, like the most frequent combination with right ventricular outflow tract obstruction (the so-called tetralogy of Fallot-type), is also morphologically very similar to the tetralogy of Fallot, but without any obstruction at the level of the right ventricular outflow tract. On the contrary, there is the possibility of a restrictive ventricular septal defect (because of the presence of hypertrophic ventriculoinfundibular fold) as well as of subaortic obstruction, either due to the prominent ventriculoinfundibular fold or to the presence of a subaortic fibrous membrane. The pathophysiology in this cases is very similar to the hemodynamics of patients with unrestrictive ventricular septal defect. Typical cardiac anomalies associated with this type of double outlet right ventricle are straddling mitral valve and aortic coarctation.

More rare are the combinations of *double outlet right ventricle with doubly committed or with noncommitted (remote) ventricular septal defect.* In patients with doubly committed ventricular septal defect the morphological characteristic is the absence (or the very severe hypoplasia) of the infundibular septum, with juxtaarterial ventricular septal defect, ventriculoarterial concordance, and frequently pulmonary stenosis. In patients with noncommitted (remote) ventricular septal defect, the defect is frequently of muscular or inlet (atrioventricular) type, and is therefore distant from both the semilunar valves, there is ventriculoarterial concordance, and pulmonary stenosis is very rare.

Associated anomalies

Dextrocardia, juxtaposition of left auricular appendages, total anomalous pulmonary venous connection, anomalous systemic venous connections, cor triatriatum, mitral stenosis, cleft of the mitral valve, common atrioventricular valve, straddling mitral and/or tricuspid valve (Fig. 2.11.5), superoinfer-

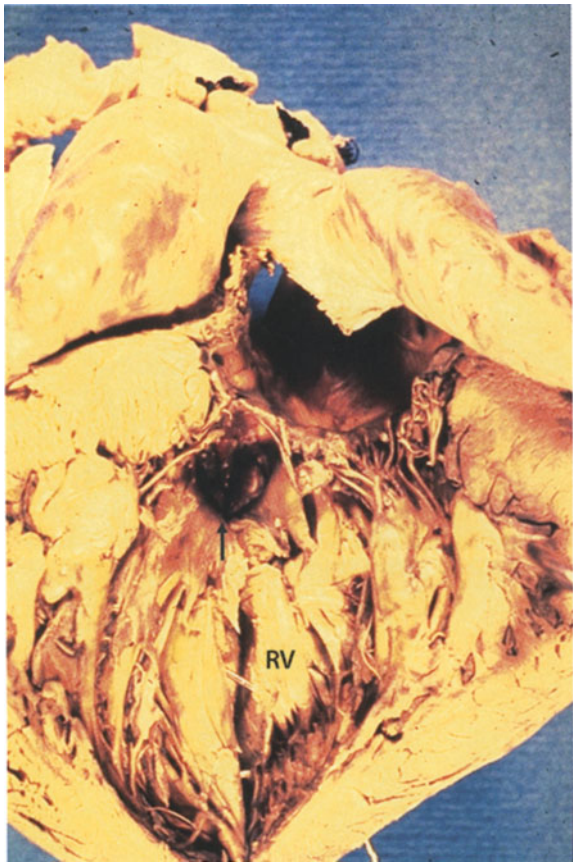

Fig. 2.11.5. Double outlet right ventricle: morphology. Heart with double outlet right ventricle, ventricular septal defect (black arrow) and straddling tricuspid valve (*RV* right ventricle) (photograph courtesy of Dr. Bruno Marino)

ior ventricles with or without criss-crossing atrioventricular connections, multiple ventricular septal defects, hypoplastic left ventricle, left ventricular outflow tract obstruction, discrete subaortic stenosis, aortic coarctation, aortic arch interruption, absent pulmonary valve, absent left pulmonary artery, and ectopia cordis have all been reported in association with double outlet right ventricle.

The origin and course of the coronary arteries are associated with the relationship between the aorta and pulmonary artery, and several variations are described, including anomalous origin of the right coronary artery from the left main coronary artery, duplication of the left anterior descending coronary artery, anomalous origin of the left anterior descending coronary artery from the right coronary artery, anomalous origin of the left circumflex artery from the right coronary artery, and single right or single left coronary artery as the most frequent anomalies.

Pathophysiology

The pathophysiology in double outlet right ventricle reflects the heterogeneity of the cardiac morphology in this abnormal ventriculoarterial connection, as well as the severity of the associated lesions. Patients with tetralogy-type of morphology, like in tetralogy of Fallot may present early in the neonatal period with right-to-left shunt or beyond infancy with relatively well-balanced circulation, depending upon the degree of obstruction to the pulmonary blood flow. Patients with double outlet right ventricle and subaortic ventricular septal defect present with large left-to-right intracardiac shunt, like patients with isolated unrestrictive ventricular septal defect. In these infants the presence of an associated left ventricular outflow tract obstruction and/or aortic coarctation further increases the left-to-right intracardiac shunt. On the contrary, substantial right-to-left intracardiac shunt is present from the first few days of life in patients with double outlet right ventricle and subpulmonary ventricular septal defect.

Diagnosis

Clinical pattern: the clinical presentation reflects the underlying morphology; cyanosis and/or heart failure can be present depending upon the type of ventriculo-arterial connection and the presence and degree of obstruction to the pulmonary and systemic blood flow, and the subsequent intracardiac streaming; the physical signs will vary according to the pathophysiologic pattern.

Electrocardiogram: right axis deviation, right ventricular hypertrophy, often with a qR pattern in the right precordial leads, more rarely biventricular hypertrophy.

Chest X-ray: it is not diagnostic and not specific, since the cardiac silhouette can vary from the aspect typical of tetralogy of Fallot to the appearance present in transposition of the great arteries.

Echocardiogram: the combination of parasternal short- and long-axis views allows complete diagnosis, with identification of the type of intracardiac morphology; subcostal and apical 4-chamber views demonstrate the precise spatial relationship between the aorta and pulmonary artery and to the ventricular septal defect, and the presence and degree of

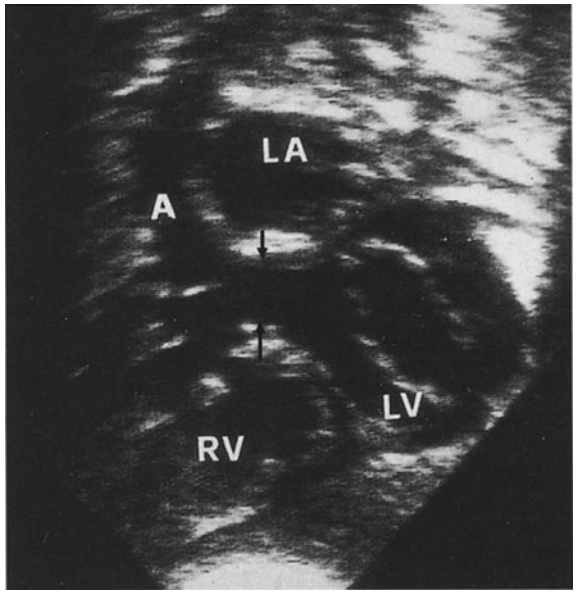

Fig. 2.11.6. Double outlet right ventricle: echocardiography with left oblique subxiphoid view showing double outlet right ventricle with ventriculoarterial concordance and unrestrictive subaortic ventricular septal defect (black arrows) (*A* aorta, *LA* left atrium, *LV* left ventricle, *RV* right ventricle) (photograph courtesy of Dr. Bruno Marino)

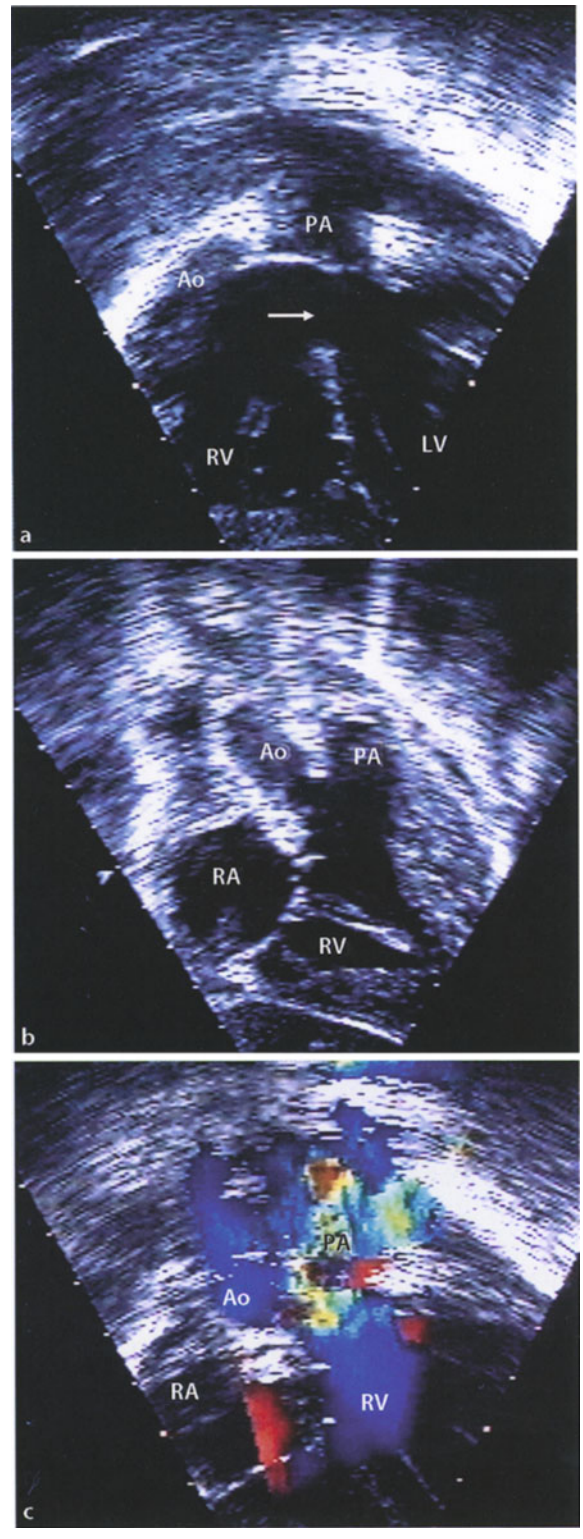

Fig. 2.11.7. Double outlet right ventricle: echocardiography in a child with Taussig-Bing, intact atrial septum, hypoplastic aortic arch, patent ductus arteriosus, and systemic pulmonary hypertension, with **a** the left oblique subxiphoid view showing the unrestrictive subpulmonary ventricular septal defect (white arrow) and **b** the long-axis subxiphoid view showing the double outlet from the right ventricle, **c** color Doppler echocardiography in the same patient of **a** and **b** showing the unrestricted double outlet from the right ventricle (*Ao* aorta, *PA* pulmonary artery, *RA* right atrium, *RV* right ventricle)

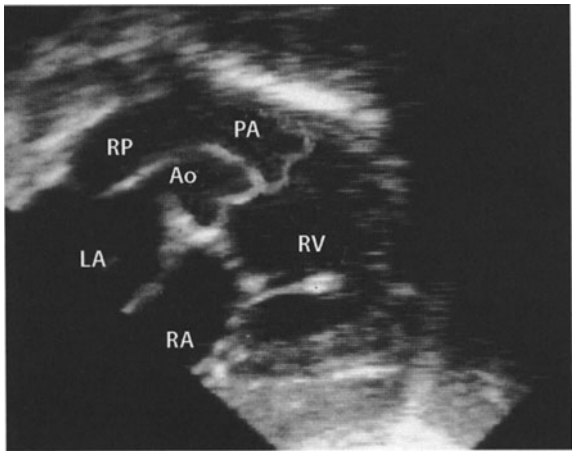

Fig. 2.11.8. Double outlet right ventricle: echocardiography in the right oblique subxiphoid view showing a double outlet right ventricle with doubly committed ventricular septal defect (*Ao* aorta, *LA* left atrium, *PA* pulmonary artery, *RA* right atrium, *RPA* right pulmonary artery, *RV* right ventricle) (photograph courtesy of Dr. Michael Rigby)

obstruction to the right and left ventricular outflow tract (Figs. 2.11.6–2.11.8);

Cardiac catheterization: this procedure is indicated in neonates with ventriculoarterial discordance and restrictive interatrial communication, where balloon atrioseptostomy (=Rashkind procedure) is required, or in order to better define the origin and course of coronary arteries in view of an arterial switch operation, or to better evaluate systemic obstructions (Fig. 2.11.9); in patients with ventriculoarterial concordance it is required in order to plan primary surgical repair, while in older children with pulmonary hypertension it is used to quantitate the pulmonary vascular resistance and to rule out pulmonary vascular obstructive disease.

Indications for surgical treatment

Palliation: modified Blalock-Taussig shunt (see chapter "Tetralogy of Fallot") can be considered in neonates with *double outlet right ventricle with concordant ventriculoarterial connection, subaortic ventricular septal defect and obstruction to the pulmonary outflow tract (so-called tetralogy of Fallot-type)* in the presence of severe cyanosis in the first days or weeks of life, when primary repair is judged to carry a higher risk than two-stage surgical treatment.

Pulmonary artery banding is considered for *double outlet right ventricle with concordant ventriculoarterial connection and subaortic ventricular septal defect, without obstruction to the pulmonary outflow tract* when the patient comes to observation after late referral with severe pulmonary hypertension (Fig. 2.11.10), in order to allow for a period with reduced pulmonary artery pressure before intracardiac repair.

Repair: according to each different type of morphology, several surgical techniques have been reported, referring to one of the following principles:

- *intraventricular repair* connecting the left ventricle to the aorta and the right ventricle to the pulmonary artery,
- *arterial switch* operation with closure of the ventricular septal defect by a prosthetic patch connecting the left ventricle to the neoaorta,
- *univentricular type of repair*, with end-to-side superior vena cava to pulmonary artery anastomosis (=bidirectional Glenn) followed by total cavopulmonary connection (=modified Fontan procedure).

Because the large variability of intracardiac morphologies, with various associated cardiac lesions, and the different surgical experiences, several alternative surgical options are available for each patient, making the choice of the operation something unique for the mismatch of the individual patient with the individual surgeon.

Surgical treatment (on cardiopulmonary bypass)

Intraventricular repair: there are different types of intraventricular repair. In all of them the left ventricle is connected with the

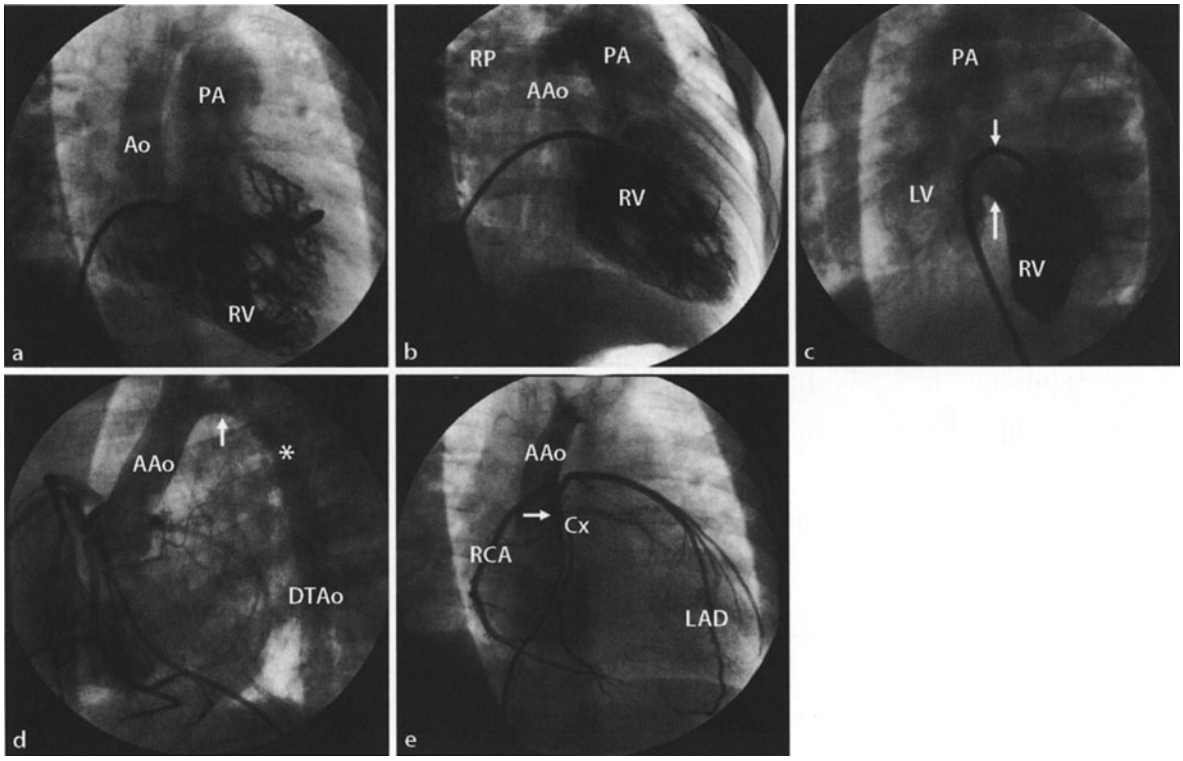

Fig. 2.11.9. Double outlet right ventricle: angiography. **a** right ventricular injection with left anterior oblique view showing the double outlet right ventricle, with hypertrophic right ventricle, small ascending aorta, very dilated main pulmonary artery, in a child with Taussig-Bing malformation, large subpulmonary ventricular septal defect, hypoplastic aortic arch, aortic coarctation and anomalous single origin of the coronary arteries from the left sinus (*Ao* aorta, *PA* pulmonary artery, *RV* right ventricle), **b** right ventricular injection in the same patient with left anterior oblique view showing the double outlet right ventricle, with small ascending aorta and very dilated main pulmonary artery (*AAo* ascending aorta, *RPA* right pulmonary artery), **c** right ventricular injection in the same patient with lateral view showing the double outlet right ventricle and the large subpulmonary ventricular septal defect (white arrows) (*LV* left ventricle), **d** aortic root injection in the same patient with lateral view showing the hypoplastic aortic arch (white arrow) with aortic coarctation (white asterisk) (*DTAo* descending thoracic aorta), **e** selective coronary artery injection in the same patient with anteroposterior view showing the single origin of the coronary arteries from the left sinus (white arrow). The patient underwent successful repair with closure of ventricular septal defect, reconstruction of the aortic arch and isthmus with an autologous patch of the native pulmonary artery, and biological valved conduit implantation between the right ventricle and pulmonary artery (*Cx* circumflex coronary artery, *LAD* left anterior descending coronary artery, *RCA* right coronary artery)

aorta, and the right ventricle with the pulmonary artery, either directly or with the interposition of a conduit.

Intraventricular tunnel repair. This type of repair, suitable for the more simple type of *double outlet right ventricle with concordant ventriculoarterial connection and subaortic ventricular septal defect*, can be performed either through right atriotomy (rarely) or right ventriculotomy. Right ventriculotomy allows easier evaluation of the adequate shape and positioning of the prosthetic patch needed to connect the left ventricle with the aorta. The ventricular size and position, combined with the relationship between the diameter of the aortic valve and the distance between the tricuspid and pulmonary valve, dictate the need for enlargement of the tunnel between left ventricle and aorta. When needed, the ventricular septal defect is enlarged by an anterior incision, with muscular resection. The prosthetic material used to create the intraventricular tunnel is generally a prosthetic tubular prosthesis (PTFE, Dacron) about 20% larger than the aortic diameter, cut at a length

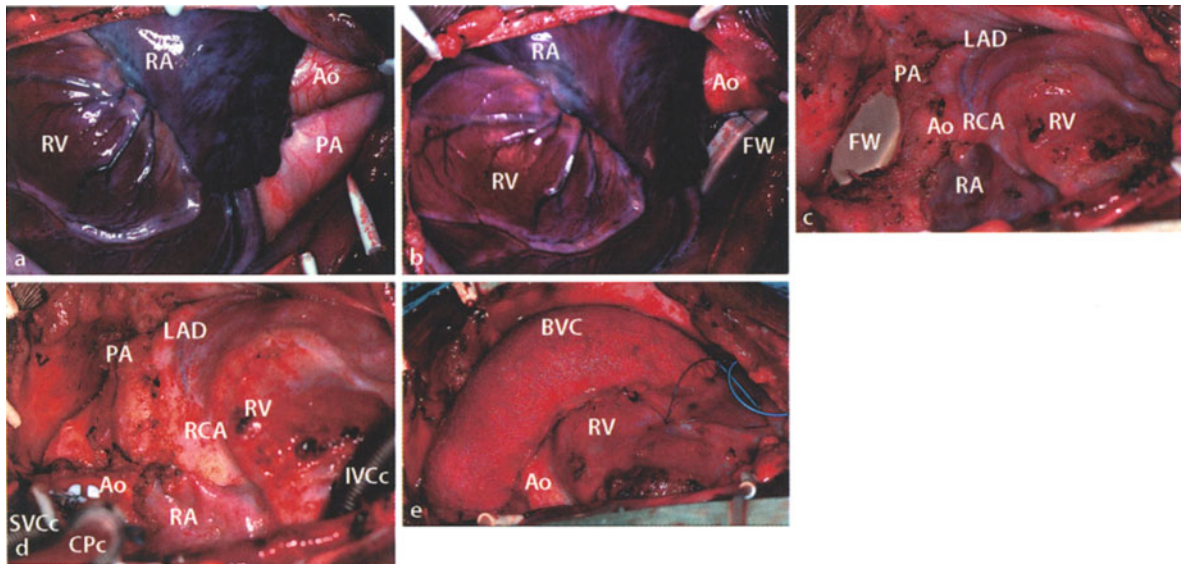

Fig. 2.11.10. Double outlet right ventricle: surgery. Intraoperative photograph in a child **a** with double outlet right ventricle, uncommitted ventricular septal defect, straddling tricuspid valve, anomalous left anterior descending coronary artery from the right coronary artery, and severe pulmonary hypertension, who underwent pulmonary artery banding with the adjustable FloWatch-PAB^{TM} at three years of age with a very dilated pulmonary artery because of severe pulmonary hypertension due to late referral (*Ao* aorta, *PA* pulmonary artery, *RA* right atrium, *RV* right ventricle), **b** after palliative surgery: pulmonary artery banding with adjustable device (*FW* FloWatch-PAB^{TM}), **c** the dissection of the FloWatch-PAB^{TM} at the time of debanding with intracardiac repair (*RCA* right coronary artery), **d** after preparation of cardiopulmonary bypass for intracardiac repair and removal of the FloWatch-PAB^{TM}, with spontaneous dilatation of the pulmonary artery (*CPc* cardioplegia cannula, *IVCc* inferior vena cava cannula, *SVC* superior vena cava cannula), **e** after intracardiac repair with implantation of a biological valved conduit (Contegra, Medtronic) between the right ventricle and pulmonary artery (*BVC* biological valved conduit)

equal to the distance between the anterior edge of the ventricular septal defect and the aortic annulus. Two-thirds of the entire circumference of the tubular prosthesis is used, in a manner to leaving an unobstructed left ventricle to aorta tunnel, but at the same time avoiding the potential bulging of the patch to obstruct the right ventricular outflow tract (Fig. 2.11.11).

In the presence of severe *obstruction to the pulmonary outflow tract*, a transannular patch or a biological valved conduit implanted between the right ventriculotomy and the pulmonary artery is used to relieve the obstruction. An alternative option is the Lecompte procedure (see below for details).

In *double outlet right ventricle with discordant ventriculoarterial connection and subpulmonary ventricular septal defect, without obstruction to the pulmonary outflow tract (so-called Taussig-Bing malformation)*, the intraventricular tunnel is more complicated

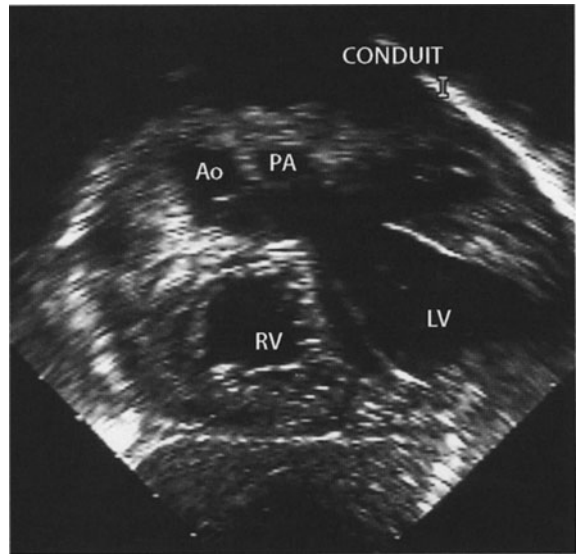

Fig. 2.11.11. Double outlet right ventricle: surgery. Postoperative echocardiography of the same patient as in Fig. 2.11.7 showing the relationship between the tunnel left ventricle-aorta and the biological valved conduit implanted between the right ventricle and pulmonary artery (*Ao* aorta, *LV* left ventricle, *PA* pulmonary artery, *RV* right ventricle)

to design and accomplished. In the *Kawashima technique*, the intraventricular tunnel is positioned posteriorly to the orifice of the pulmonary valve, if there is enough distance between the tricuspid and pulmonary valves. If the distance between the tricuspid and pulmonary valves is inadequate, the intraventricular tunnel has to remain anterior to the orifice of the pulmonary valve. In both cases the ventricular septal defect needs to be enlarged by anterior resection of the infundibular septum.

In patients with *double outlet right ventricle with discordant ventriculoarterial connection and obstruction to the pulmonary outflow tract* the best surgical option is the *Lecompte procedure (or REV = Réparation à l'Etage Ventriculaire)* (see chapter "Transposition of the great arteries"); after right ventriculotomy and infundibular resection, the left ventricle is connected to the aorta by closure of ventricular septal defect with a straight patch; after transection and shortening of the ascending aorta and transfer of the pulmonary artery bifurcation anterior to the aorta (*Lecompte maneuver*), the right ventricle-to-pulmonary artery continuity is obtained with reimplantation of the transected pulmonary artery directly on the right ventricle for its posterior wall, while the anterior aspect is connected to the rest of the right ventriculotomy with a monocusp pericardial patch.

- **Arterial switch operation:** the crucial point is the surgical approach for closure of the ventricular septal defect. The possibilities are through right atriotomy (ideal for perimembranous and inlet defects), through the aortic valve (neopulmonary valve) after resection of the aortic buttons with the orifices of the coronary arteries (ideal for outlet types of subaortic defects), or through the pulmonary valve (neoaortic valve) (ideal for subpulmonary type of defects). The rest of the procedure is like that for the conventional arterial switch (see chapter "Transposition of the great arteries").

Since the most frequent complications after arterial switch for Taussig-Bing type of double outlet right ventricle are residual or recurrent right ventricular outflow tract obstruction and residual or recurrent neoaortic valve regurgitation, the following technical steps are important to follow:

- avoid pulmonary artery banding and perform whenever possible a one-stage repair;
- prepare the pulmonary arteries with very extended mobilization, including the pulmonary artery branches;
- perform a radical muscle resection in the presence of hypertrophic infundibular septum;
- avoid closure of the ventricular septal defect through the aortic valve;
- always perform the *Lecompte maneuver*;
- reduce the distortion of the neoaortic valve during coronary arteries transfer, either using the trap-door or punch technique;
- avoid a discrepancy between the size of the neoaortic root and the ascending aorta;
- use fresh autologous pericardial patch(es) for neopulmonary artery reconstruction.

An alternative surgical technique is the *Nikaidoh aortic translocation*, where the aortic root, including aortic valve and coronary arteries is isolated from the right ventricle, the pulmonary artery is transected and the area between the ventricular septal defect and the proximal stump of the transected pulmonary artery is widely incised; the left ventricle is connected to the aorta with a patch roofing the opened ventricular septal defect, while the right ventricle is connected to the transected pulmonary artery like in the Lecompte procedure.

In the Taussig-Bing type of double outlet right ventricle with aortic arch obstruction, a surgical technique recently proposed (Dr. Sano) consists of the transection of the great arteries, ductus arteriosus, descending thoracic aorta, and aortic arch with aortotomy incision from the aortic arch to the distal ascending aorta, followed by creation of an aortopulmonary window, anastomosis of the descending thoracic aorta to the posterior wall of the aortic arch, anastomosis of the neoaorta to the aortic arch with rerouting of

the coronary artery (without need for coronary arteries reimplantation), and reconstruction of the neo-right ventricular outflow tract after the Lecompte maneuver.

Univentricular type of repair: in the presence of complicated intracardiac anatomy like the presence of inlet type of noncommitted ventricular septal defect with straddling tricuspid valve, common atrioventricular valve, or hypoplasia of the right ventricular chamber, the biventricular is either not feasible or is associated with too high of a risk. In these patients the univentricular type of repair is the preferred surgical option. In these cases, the first step is a bidirectional Glenn, followed by the modified Fontan procedure (see chapter "Single ventricle"), preceded or not by pulmonary artery banding, depending upon the presence or absence of obstruction to the pulmonary blood flow.

Potential complications

Intraventricular repair: potential complications after intraventricular repair include residual or recurrent ventricular septal defect, residual or recurrent right and/or left ventricular outflow tract obstruction, arrhythmias, complete atrioventricular block, atrioventricular valve regurgitation.

Arterial switch operation: the most frequent complications are residual or recurrent right ventricular outflow tract obstruction, and residual or recurrent neoaortic valve regurgitation.

Univentricular type of repair: see chapter "Single ventricle".

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Chapter 2.12 Double discordance

Incidence

Double discordance (atrioventricular and ventriculo-arterial discordance), otherwise called "congenitally corrected transposition of the great arteries", is the 17th most common congenital heart defect (0.5–1.4% of all congenital heart defects).

Morphology

In this fascinating cardiac malformation the morphological left atrium (= pulmonary venous atrium) is connected via a tricuspid valve with the morphological right ventricle, from which the aorta originates, while the morphological right atrium (= systemic venous atrium) is connected via a mitral valve with the morphological left ventricle, from which originates the pulmonary artery (Figs. 2.12.1–2.12.4). Therefore two discordant connections, atrioventricular and ventriculoarterial (= double discordance), occur in sequence on each side of the heart. The classical definition of "congenitally corrected transposition of the great arteries" derived from the observation that the effects of transposition of the great arteries are "corrected" by the congenital inversion of the two ventricles, with the two circulatory pathways "physiologically" in series, despite the anatomic derangements.

The four chambers of the heart have distinct features, which identify them regardless of their actual connection or spatial location. Therefore, even if a ventricle is located on the right side, it can be identified as a morphologic left ventricle.

The right atrial appendage is identified by the broad triangular shape as opposed to the narrow fingerlike left atrial appendage. The right ventricle is identified by the presence of the moderator band, muscle tissue that traverses the ventricle horizontally near the apex; the tricuspid valve, always connected to the right ventricle, has multiple papillary muscle attachments to the septum (while the mitral valve has none), and it is separated from the

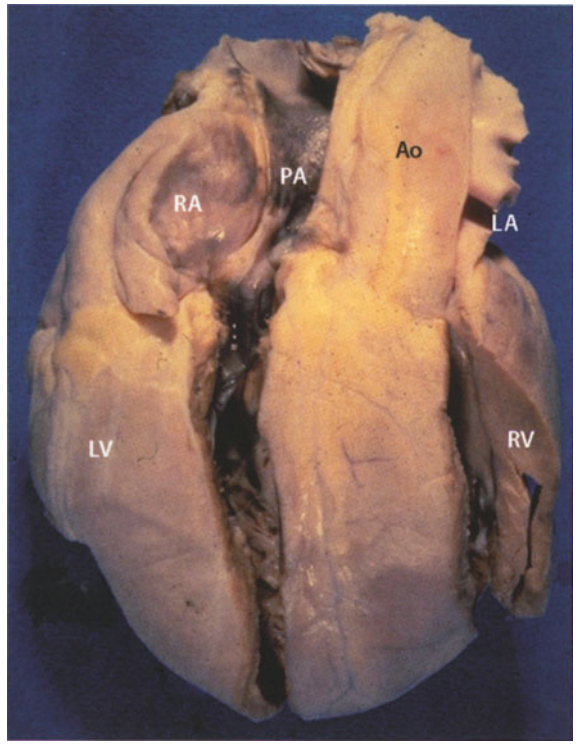

Fig. 2.12.1. Double discordance: morphology. External appearance of a heart with atrioventricular and ventricular discordance (*Ao* aorta, *LA* left atrium, *LV* left ventricle, *PA* pulmonary artery, *RA* right atrium, *RV* right ventricle) (photograph courtesy of Dr. Marino

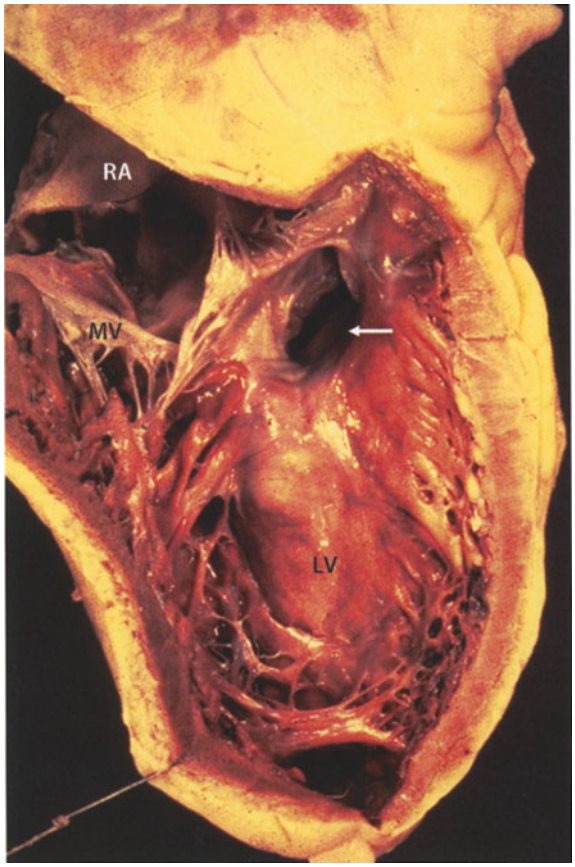

Fig. 2.12.2. Double discordance: morphology. Appearance of a heart with atrioventricular discordance with a large ventricular septal defect (white arrow) (*LV* left ventricle, *MV* mitral valve, *RA* right atrium) (photograph courtesy of Dr. Marino)

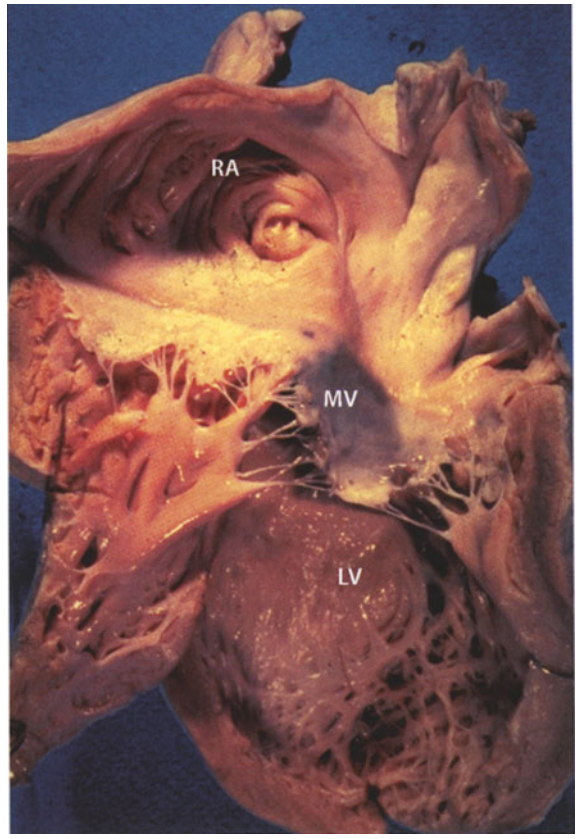

Fig. 2.12.3. Double discordance: morphology. Appearance of a heart with atrioventricular discordance (*LV* left ventricle, *MV* mitral valve, *RA* right atrium) (photograph courtesy of Dr. Marino)

semilunar (generally the pulmonary, but in these hearts the aortic) valve by a muscular band called crista supraventricularis or conus, creating a well-defined right ventricular infundibulum. The mitral valve, always connected to the left ventricle, is in fibrous continuity with the semilunar (generally the aortic, but in these hearts the pulmonary) valve.

In these hearts an atrial situs solitus or inversus can be present, as well as supero-inferior disposition of the ventricular chambers can occur, with a more or less horizontal interventricular septum. Dextrocardia is present in 25% of the cases.

In hearts with double discordance with atrial situs solitus, there are two atrioventricular nodes: the first is in normal position, at the apex of the triangle of Koch in front of the orifice of the coronary sinus; the second is located close to the orifice of the right atrioventricular valve beneath the ostium of the right auricular appendage. The main important point is that, because of the septal malalignment, the penetrating bundle of His generally arises from the second atrioventricular node, penetrating across the edge of the muscular septum to reach the left-sided right ventricular septal surface.

The coronary arteries originate from the facing sinuses of the aortic valve, generally with a mirror-image distribution, following the appropriate ventricle.

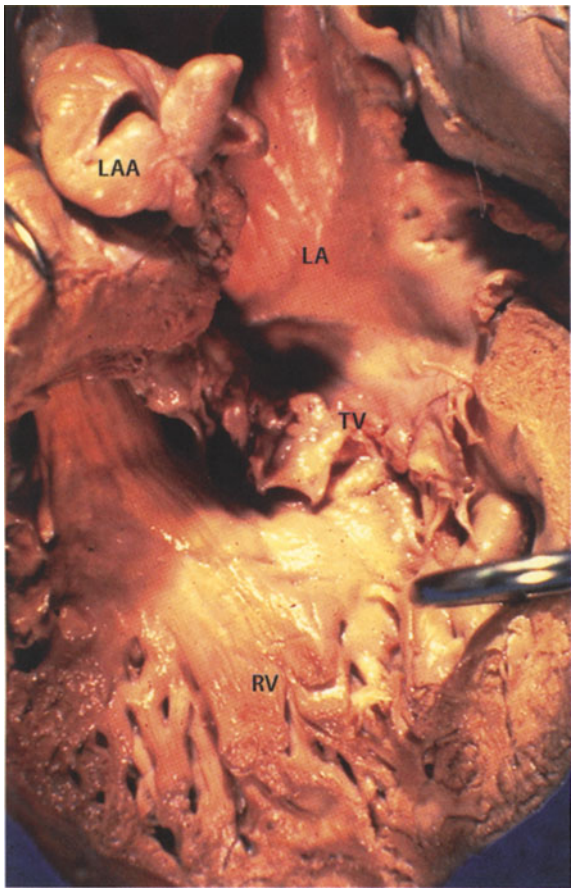

Fig. 2.12.4. Double discordance: morphology. Appearance of a heart with atrioventricular discordance (*LAA* left auricular appendage, *LA* left auricular atrium, *RV* right ventricle, *TV* tricupid valve) (photograph courtesy of Dr. Marino)

Associated anomalies

In double discordance, probably fewer than 1% of individuals have no associated malformations.

The most frequent and most important associated lesions are ventricular septal defect (up to 80% of cases), tricuspid valve anomalies, corresponding to abnormalities of the systemic atrioventricular valve (in up to 60% of cases), pulmonary stenosis or atresia (30–50%), dextrocardia (25%) and complete atrioventricular block (12–33% of cases).

The ventricular septal defect is most frequently perimembranous, in the subpulmonary position, generally unrestrictive because of the malalignment between the atrial and ventricular septa. In 10% of cases, more frequently in Asian patients, the ventricular septal defect is subarterial and in proximity of both semilunar valves.

Obstruction to the outflow tract of the morphologically left ventricle (=pulmonary stenosis) is uncommon as an isolated associated lesion, because it almost always is associated with a ventricular septal defect; the obstruction can be valvular and/or subvalvular (more frequently) and fixed and/or dynamic (more frequently) and can be caused by muscular tissue (wedging of the subpulmonary outflow tract between the infundibular septum and the free ventricular wall) due to the oblique orientation of the long axis of the pulmonary outflow tract from the right-sided left ventricle, by aneurysmal bulging of fibrous tissue derived from the membranous septum, by fibrous tissue tags derived either from the atrioventricular valve or from a thickened and fused pulmonary valve, occasionally bicuspid.

Tricuspid valve (is the valve functioning as the systemic atrioventricular valve) anomalies, very frequent (up to 90% of cases in autopsy series), include dysplasia with or without displacement of the septal and posterior leaflets (=Ebstein-like malformation), straddling or overriding an inlet muscular ventricular septal defect. Mitral valve anomalies are also quite frequent (up to 55% of cases). Straddling of an atrioventricular valve can be associated with the hypoplasia of the ipsilateral ventricle.

Complete atrioventricular block is quite peculiar in this malformation, being reported in already 10% of newborns and with a constant progression in the natural history of the disease, with 2% of patients per year affect after establishment of the diagnosis of double discordance. Additional rhythm disturbances have been reported: Wolff-Parkinson-White syndrome, supraventricular tachycardia, atrial flutter and fibrillation.

Coronary artery anomalies potentially complicating anatomical repair have been reported in 45% of patients, including single coronary artery orifice (the most frequent

coronary artery anomaly, mostly originating from the right-facing sinus, but reported also from the non-facing sinus), left anterior coronary artery originating from the right coronary artery, and eccentric coronary orifices. Hypoplasia of one of the two ventricles generally occurs in the presence of a straddling atrioventricular valve (ipsilateral to the hypoplastic ventricle).

Less frequently have been reported situs inversus (with mirror-image relation), supravalvular left atrial ring, atrial septal defect, complete atrioventricular septal defect, straddling mitral valve, double outlet right ventricle, discrete subvalvular or valvular aortic stenosis, patent ductus arteriosus, aortic coarctation, aortic arch interruption, aortic atresia, pulmonary atresia with intact ventricular septum, coarctation of the left pulmonary artery.

By definition these hearts cannot present with ambiguous atrial situs (left or right atrial isomerism).

The isolated atrioventricular discordance, without ventriculoarterial discordance (also called isolated ventricular inversion), has rarely been observed, generally with associated ventricular septal defect.

Pathophysiology

The two discordant connections occur in a sequence ensuring that the blood flow continues in its usual physiologic pathway, with the oxygenated blood coming from the left atrium which reaches the aorta after passing through the right ventricle, and the desaturated blood coming from the right atrium which reaches the pulmonary artery after passing through the left ventricle. As a consequence, the oxygen saturations in the heart chambers and in the great arteries are normal, even if the blood flows through the wrong atrioventricular valves and ventricles.

Patients with double discordance generally came to observation as a result of the presence, type and severity of the associated lesions. According with the associated lesions patients can present with cyanosis (patients with pulmonary stenosis or atresia), heart failure (patients with ventricular septal defect without pulmonary stenosis or atresia and/or with tricuspid valve regurgitation corresponding to regurgitation of the systemic atrioventricular valve), a combination of cyanosis and heart failure, or bradycardia due to the congenital complete atrioventricular block.

Diagnosis

Clinical pattern: patients with isolated double discordance may remain asymptomatic through adulthood; rhythm disturbances and tricuspid valve regurgitation are more frequent after the third or fourth decades of life; congestive heart failure, due to impaired right (systemic) ventricular function, develops in the majority (66%) of patients older than 50 years; most patients with associated lesions present in infancy with a murmur or heart failure; older children may present with growth failure and exercise intolerance; bradycardia subsequent to complete atrioventricular block can occur at any age, with or without heart failure; cyanosis is present only in patients with ventricular septal defect and severe pulmonary stenosis or atresia; tracheal compression has been reported in the presence of a descending thoracic aorta located on the contralateral side of the ascending aorta.

Electrocardiogram: it may provide the most significant clue of this malformation; presence of Q waves in the right precordial leads with absent Q waves over the left precordial leads, in the absence of the criteria for right ventricular hypertrophy; congenital or developing complete atrioventricular block is also suggestive of double discordance.

Chest X-ray: the anteroposterior chest X-ray shows the characteristic features, including the convex prominence of the upper left heart border with simultaneous mild convexity in the anticipated position of the main pulmonary

artery, due to the side-by-side arrangement of the great arteries, with the levo-positioned ascending aorta originating from the left-sided morphological right ventricle (=ascending aortic shadow); cardiomegaly is present in the presence of large ventricular septal defect and/or severe tricuspid valve regurgitation.

Echocardiogram: (Figs. 2.12.5–2.12.12) the subcostal view allows identification of the interventricular septum, along with the atrioventricular valves and the interatrial septum; the recognition of the malalignment between the atrial and ventricular septum and the morphology of the two atrioventricular valves allows the diagnosis of atrioventricular discordance; the subcostal view also shows the presence of ventricular hypoplasia, as well as the pulmonary outflow tract; Doppler echocardiography provides accurate information about the function of the atrioventricular valves and on the presence and degree of outflow tract obstruction.

Cardiac catheterization: very rarely required to perform the diagnosis; it is useful to provide the hemodynamic data, particularly in the presence of ventricular septal defect (Fig. 2.12.13), and to confirm the origin and distribution of the coronary arteries; it carries a substantial risk of inducing a complete atrioventricular block during the procedure because the atrioventricular bundle is located on the left ventricular side of the septum, and the morphological left ventricle is connected with the right atrium.

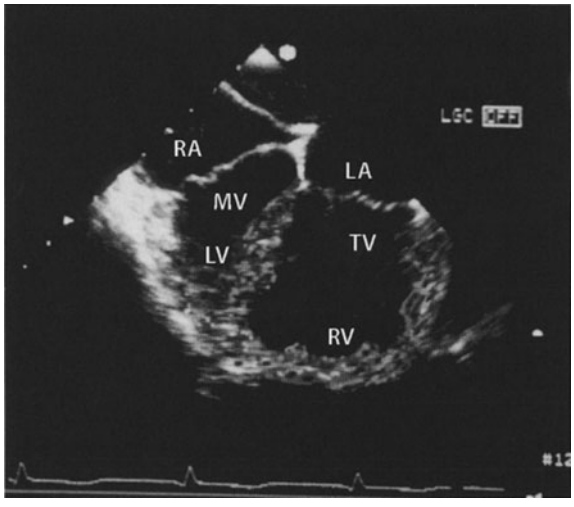

Fig. 2.12.5. Double discordance: echocardiography. The 4-chamber view showing the atrioventricular discordance (*LA* left atrium, *LV* left ventricle, *MV* mitral valve, *RA* right atrium, *RV* right ventricle, *TV* tricuspid valve) (photograph courtesy of Dr. Michael Rigby)

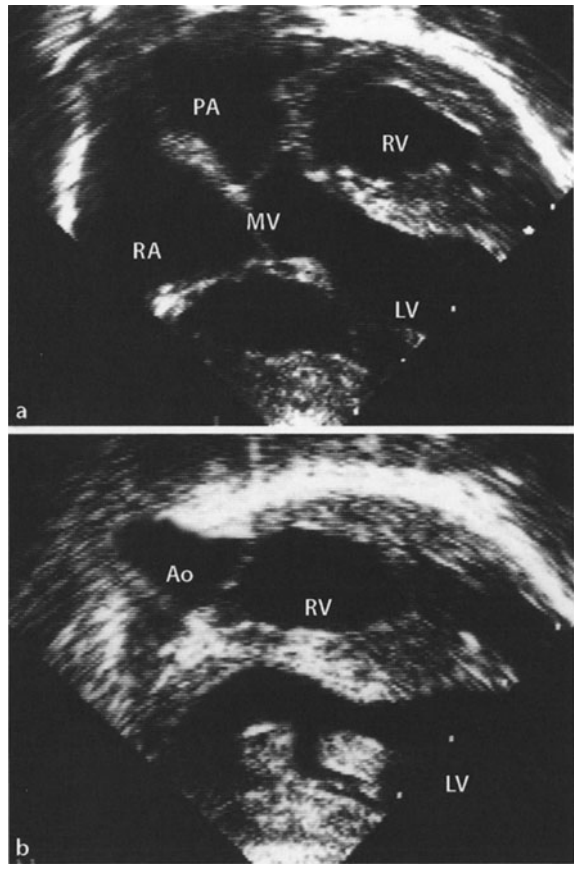

Fig. 2.12.6. Double discordance: echocardiography. Subcostal view (**a**) showing the atrioventricular and ventriculoarterial discordance (*LV* left ventricle, *MV* mitral valve, *PA* pulmonary artery, *RA* right atrium, *RV* right ventricle) and **b** subcostal view in the same patient confirming the ventriculoarterial discordance (*Ao* aorta) (photographs courtesy of Dr. Michael Rigby)

Indications for surgical treatment

No surgical treatment is required for the rare patient without associated anomalies, since their life expectancy has been reported to be near normal. The surgical management

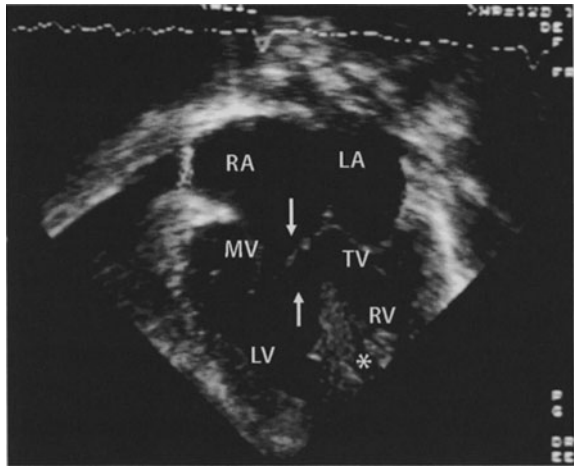

Fig. 2.12.7. Double discordance: echocardiography with 4-chamber view showing the atrioventricular discordance and a large ventricular septal defect (white arrows), with the moderator band (white asterisk) identifying the right ventricle (*LA* left atrium, *LV* left ventricle, *MV* mitral valve, *RA* right atrium, *RV* right ventricle, *TV* tricuspid valve) (photograph courtesy of Dr. Michael Rigby)

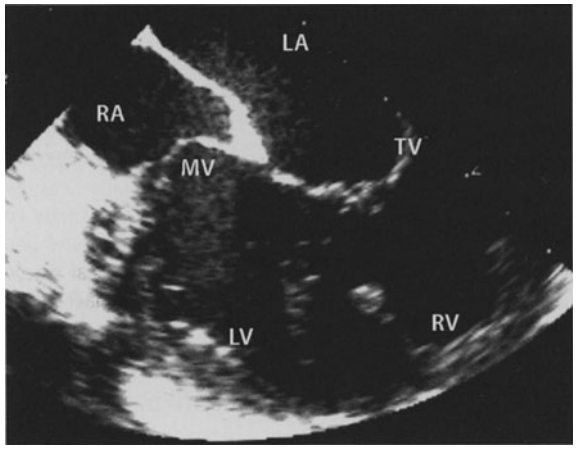

Fig. 2.12.8. Double discordance: transesophageal echocardiography showing the atrioventricular discordance (*LA* left atrium, *LV* left ventricle, *MV* mitral valve, *RA* right atrium, *RV* right ventricle, *TV* tricuspid valve) (photograph courtesy of Dr. Pierre-Guy Chassot)

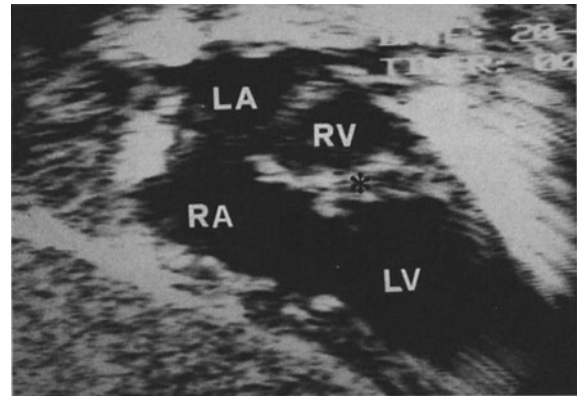

Fig. 2.12.9. Double discordance: echocardiography with 4-chamber subxiphoid view showing the superoinferior relationship of the two ventricles, with an horizontal interventricular septum (black asterisk) and the two atrioventricular valves in a parallel relationship, in the presence of atrioventricular discordance (*LA* left atrium, *LV* left ventricle, *RA* right atrium, *RV* right ventricle) (reproduced with permission from Marino B, Thiene G (1990) Atlante di anatomia ecocardiografica delle cardiopatie congenite, USES, Firenze

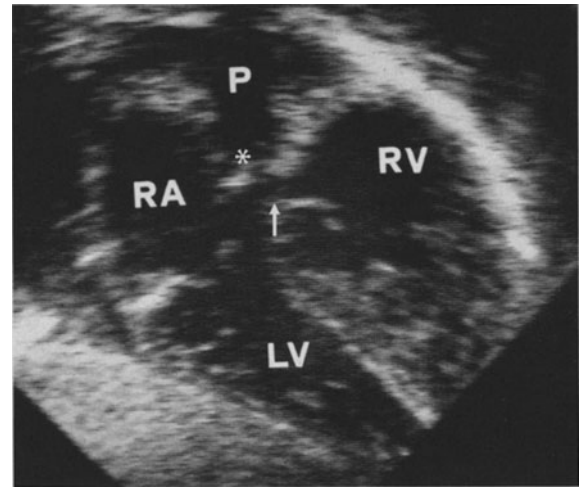

Fig. 2.12.10. Double discordance: echocardiography with right oblique subxiphoid view showing the double atrioventricular discordance with ventricular septal defect (white arrow) and the subpulmonary stenosis (white asterisk) (*LV* left ventricle, *P* pulmonary artery, *RA* right atrium, *RV* right ventricle) (reproduced with permission from Marino B, Thiene G (1990) Atlante di anatomia ecocardiografica delle cardiopatie congenite, USES, Firenze)

of even simple associated defects, such a ventricular septal defect or pulmonary stenosis, has been reported to be associated with much higher mortality and morbidity rates in these patients than in patients with an otherwise normal heart. From a technical point of view, the surgical approach to address the ventricular septal defect or the pulmonary stenosis is difficult, and the risk of inducing a complete atrioventricular block is high. Furthermore, despite technical success, the above procedures may not result in functional improvement.

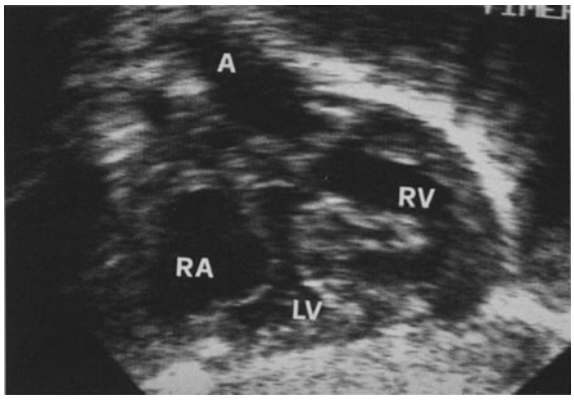

Fig. 2.12.11. Double discordance: echocardiography with subxiphoid view showing the double atrioventricular discordance; note that with a minimal inclination of the ultrasound probe compared with Fig. 2.12.10 it is possible to simultaneously visualize the right atrioventricular connection and the left ventriculoarterial connection (*A* aorta, *LV* left ventricle, *RA* right atrium, *RV* right ventricle) (reproduced with permission from Marino B, Thiene G (1990) Atlante di anatomia ecocardiografica delle cardiopatie congenite, USES, Firenze)

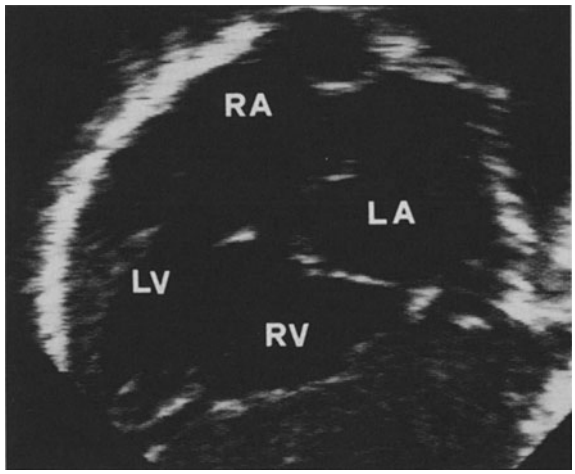

Fig. 2.12.12. Double discordance: echocardiography with 4-chamber view showing the atrioventricular discordance in a patient with dextrocardia (*LA* left atrium, *LV* left ventricle, *RA* right atrium, *RV* right ventricle) (photograph courtesy of Dr. Marino)

Palliations: the initial surgical approach in infancy can be a palliative procedure, including pulmonary artery banding in the presence of a large ventricular septal defect with pulmonary hypertension, or a modified Blalock-Taussig shunt (see chapter "Tetralogy of Fallot") in the presence of severe cyanosis due to pulmonary stenosis.

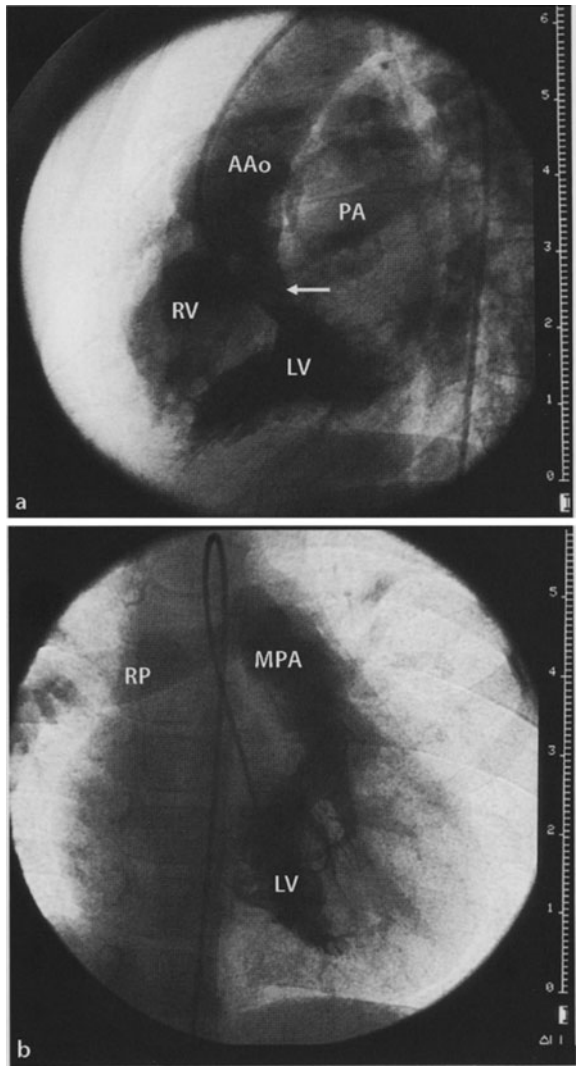

Fig. 2.12.13. Double discordance: angiography. **a** Left anterior oblique projection with contrast injection into the morphological left ventricle reached retrogradly from the aorta, showing the ventriculoarterial discordance and a ventricular septal defect (white arrow) in a patient with double discordance, bilateral superior vena cava and straddling tricuspid valve (*AAo* ascending aorta, *LV* left ventricle, *PA* pulmonary artery, *RV* right ventricle), **b** anteroposterior projection in the same patient with contrast injection into the morphological left ventricle reached retrogradly from the aorta, showing the ventriculoarterial discordance, the relative hypoplasia of the left ventricle, and a subpulmonary obstruction (*MPA* main pulmonary artery, *RPA* right pulmonary artery)

Conventional repair: depending on the age at presentation and of the specific combination of the associated lesions, a conventional surgical repair of double discordance may involve tricuspid valve repair or replacement,

closure of the ventricular septal defect, implant of a biological valved conduit between the left ventricle and the pulmonary artery and a pacemaker implantation. With the conventional repair the morphologically right ventricle remains the systemic ventricle, and the morphological right atrioventricular valve (tricuspid valve) remains the systemic valve.

Double (=atrial and arterial) switch procedure: because of the disappointing medium and long-term results of the conventional approach, within the last years the surgical treatment has been directed toward a repair where the morphological left ventricle and the morphological left atrioventricular valve (mitral valve) are restored to the systemic circulation.

The following conditions are required for a double switch procedure:

- absence of major atrioventricular valve straddling,
- balanced ventricular chambers,
- adequate left ventricular function and pressure (at least 75% of the systemic right ventricle), either because of the presence of an unrestrictive ventricular septal defect or because of a previous pulmonary artery banding,
- coronary arteries not precluding transfer and reimplantation.

With the double switch procedure the patients without pulmonary stenosis or atresia undergo atrial switch (Mustard or Senning procedure; see chapter "Transposition of the great arteries") and arterial switch (Jatene procedure; see chapter "Transposition of the great arteries"), with closure of ventricular septal defect when present.

Patients with left ventricular outflow tract obstruction (=pulmonary stenosis or atresia) undergo atrial switch (Mustard or Senning procedure; see chapter "Transposition of the great arteries"), closure of ventricular septal defect and implant of an extracardiac biological valved conduit between the right ventricle and the pulmonary artery (Rastelli operation; see chapter "Transposition of the great arteries") or the direct implantation of the transected pulmonary artery on the right ventriculotomy (Lecompte procedure; see chapter "Transposition of the great arteries").

The most difficult patients to treat are the patients coming to observation later in life, like adolescents or young adults, with heart failure due to dysfunctioning of the morphological right ventricle as the systemic ventricle, either during their natural history or after conventional type of repair. In these cases the double switch needs a previous period of left ventricular retraining, obtained with pulmonary artery banding. Due to the particular situation of these patients requiring late left ventricular retraining, left ventricular hypertrophy and function adequate to sustain the systemic circulation for the long term is very difficult to obtain. A progressive pulmonary artery banding with an externally adjustable device (FloWatch-R-PAB) seems to be the most promising technique. In the presence of severe heart failure, particularly of biventricular failure, heart transplant has to be taken into consideration.

One and half ventricular repair: while patients with severe hypoplasia of either ventricle (frequently with associated straddling of the ipsilateral atrioventricular valve), where a biventricular repair is not feasible, are managed by a univentricular type of repair (see chapter "Single ventricle"), in the presence of a relatively hypoplastic ventricle, the one and half ventricular repair (end-to-side anastomosis of the superior vena cava to the right pulmonary artery in addition to intracardiac repair, in order to reduce the volume overload of the small/malfunctioning right ventricle) is the procedure of choice. The surgical approach of one and half ventricular repair in double discordance treated by the double switch procedure has the advantages of eliminating the risk of superior vena cava obstruction (present with conventional atrial rerouting), leaving more intra-

atrial space available for the pulmonary venous return, volume unloading the right ventricle made smaller in the case of right ventricle to pulmonary artery implantation, reducing the suture lines on the right atrium, therefore reducing the risk for supraventricular arrhythmias, reducing the duration of myocardial ischemia thanks to a much simplified interatrial baffle (only the inferior vena cava needs to be baffled to the tricuspid valve), and reducing the flow across the right ventricle to pulmonary artery valved conduit.

Pace-maker implantation: in the presence of complete atrioventricular block in infants and children with double discordance there is indication pacemaker implantation regardless of the heart rate, for symptoms or heart failure. In the absence of complete atrioventricular block at the end of a reparative procedure, the permanent pacemaker leads are placed for future utilization.

Surgical treatment (on cardiopulmonary bypass)

Ventricular septal defect: in most cases the ventricular septal defect is perimembranous. The peculiar position of the atrioventricular nodes and penetrating bundle of His increases the risk of complete atrioventricular block during closure of the defect, even with the stitches being applied to the morphological right side of the defect in order to minimize the risk of a lesion to the conduction tissue. The surgical approach can be from a right atriotomy (through the right-sided mitral valve), an aortotomy (with access to the left-sided right ventricular aspect of the interventricular septum), a right ventriculotomy when a right ventricle to pulmonary artery is required, or from a low left ventriculotomy in conventional repair, where the morphological left ventricle remains the subpulmonary ventricle.

Tricuspid valve: tricuspid valve regurgitation is generally treated by valve repair with annuloplasty, particularly with a double switch type of approach; when the tricuspid valve remains as the systemic atrioventricular valve (like in conventional repair), a valve replacement is more frequently required.

Pulmonary stenosis: the pulmonary outflow tract obstruction is treated by implantation of an extracardiac biological valved conduit between the right ventricle and the pulmonary artery with the double switch approach, from the apex of the left ventricle to the pulmonary artery with the conventional approach. Rarely, it is possible to directly relieve the obstruction by excision of fibrous subvalvular tags or a subvalvular fibrous diaphragm, or by pulmonary valvotomy in the presence of isolated pulmonary valve stenosis.

Potential complications

Conventional repair: early potential complications are arrhythmias, complete atrioventricular block, residual ventricular septal defect, residual tricuspid valve regurgitation and residual right ventricular outflow tract obstruction. In the follow-up, a substantial percentage (up to 67%) of patients treated with conventional repair develop congestive heart failure and dysfunction with morphological right ventricular failure; this complication is strongly associated with the presence of regurgitation of the morphological right atrioventricular valve (tricuspid valve), and particularly with tricuspid valve repair or replacement.

Double (=atrial and arterial) switch procedure: myocardial failure with low cardiac output can occur after such a long and complicated type of procedure, particularly in the presence of previous surgical treatment(s). Other potential complications are the occurrence of complete atrioventricular block, residual or recurrent systemic and/or

pulmonary venous obstructions (because of the atrial rerouting), residual or recurrent atrial septal defect, residual or recurrent tricuspid valve regurgitation, residual or recurrent ventricular septal defect, residual or recurrent left or right ventricular outflow tract obstruction.

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Chapter 2.13 Straddling atrioventricular valve

Incidence

A straddling atrioventricular valve has been recognized in 0.4 to 0.7% of children with congenital heart defects.

Morphology

Straddling atrioventricular valves are defined as valves with chordal attachments into a contralateral ventricle (= the tension apparatus of the atrioventricular valve has biventricular insertions, or it is attached to both sides of the interventricular septum), and they have been classified into three types:

- type A: chordal insertion into the contralateral ventricle near the edge of the ventricular septum,
- type B: chordal insertion along the contralateral ventricular septum,
- type C: chordal insertion into the free wall and/or the papillary muscles of the contralateral ventricle.

Straddling of an atrioventricular valve must be differentiated by overriding of the atrioventricular valve annulus (Fig. 2.13.1), defined as commitment of a valve annulus to the contralateral ventricle (= the atrioventricular valve annulus is connected to ventricles on both sides of a septal structure), and classified into three types:

- minor: less than 50% of the atrioventricular annulus committed to the contralateral ventricle,
- major: about 50% of the atrioventricular annulus committed to each ventricle,

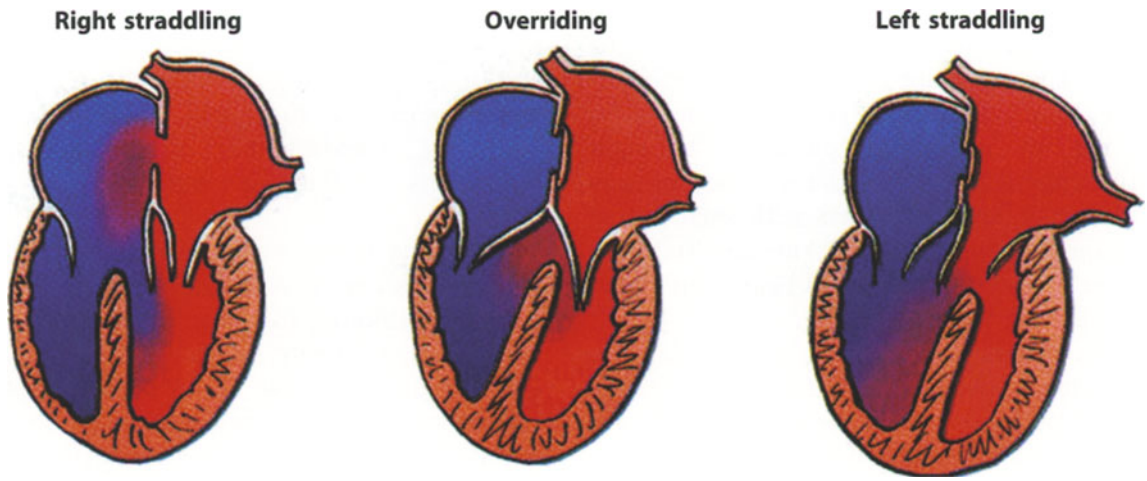

Fig. 2.13.1. Straddling atrioventricular valve: morphology. Schematic drawing showing the difference between straddling and overriding atrioventricular valve (photograph courtesy of Dr. Pierre-Guy Chassot)

A. F. Corno, *Congenital Heart Defects*

- double inlet ventricle: more than 50% of both atrioventricular valves committed to a single ventricular chamber.

By inference, both straddling and overriding of an atrioventricular valve implies the presence of a ventricular septal defect, generally a posterior defect of the inlet septum with straddling or overriding of the tricuspid valve, and an anterior defect of the outlet septum with straddling or overriding of the mitral valve. Straddling and overriding of an atrioventricular valve my coexist in the same heart, and straddling of the tricuspid valve is more frequent than straddling of the mitral valve. Because of the malalignment of the atrial septum and ventricular septum typically present with a straddling atrioventricular valve, anomalous positions of the conduction tissue are very frequent.

Straddling tricuspid valve (Fig. 2.13.2): the central feature of hearts with straddling and overriding of the tricuspid orifice is that they are intermediate between normal hearts with a concordant atrioventricular connection and those with double inlet left ventricle and right-sided rudimentary right ventricle. The disposition of the conduction tissues reflects this intermediate status, since the atrioventricular node is formed at the point at which the ventricular septum, overridden by the abnormal tricuspid orifice, makes contact with the atrioventricular junction. According to the degree of override, the node can be formed at any point around the tricuspid orifice. This arrangement is well accounted for on the basis of partial expansion of the right atrioventricular orifice across the primary ventricular septum. Hearts with such partial expansion represent an intermediate stage between the normal heart and hearts with double inlet left ventricle.

The presence of a straddling tricuspid valve is accompanied by marked malalignment of the ventricles relative to the atria, with the angle between the ventricular septum and the atrial septum in the short-axis projection averaging 60 degrees (the normal ventriculoatrial septal angle averaging 5 degrees), and the right ventricular sinus (=inflow tract) is significantly smaller than the left.

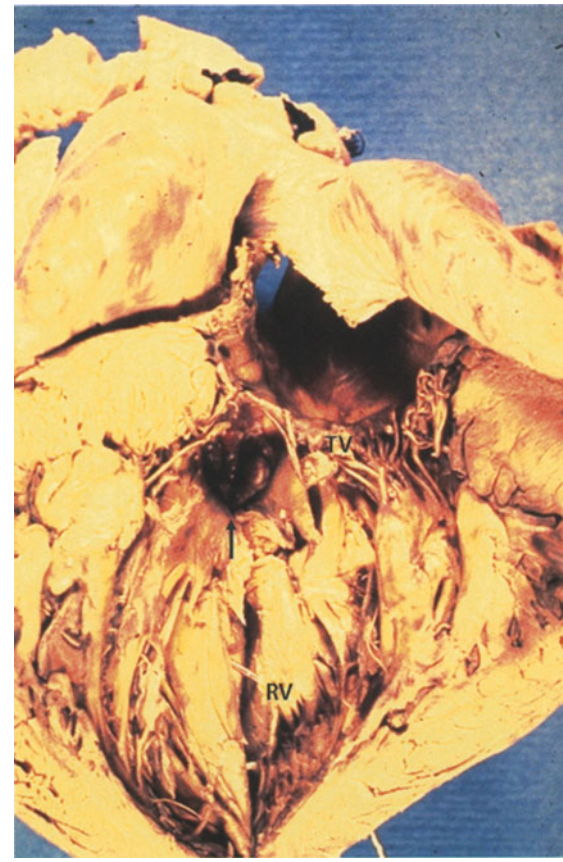

Fig. 2.13.2. Straddling tricuspid valve: morphology. Heart with double outlet right ventricle, ventricular septal defect (white arrow) and straddling tricuspid valve (*RV* right ventricle, *TV* tricuspid valve) (photograph courtesy of Dr. Bruno Marino)

The nonstraddling part of the tricuspid valve opens into the small right ventricle, while the straddling part of the tricuspid part opens into the larger left ventricle.

Straddling mitral valve (Fig. 2.13.3): wide varying anatomy is associated with the presence of straddling of the mitral valve, however, with relatively uniform morphologic features distinguished on the basis of the segmental analysis: cardiac malposition associated with hypoplasia of the morphological right ventricle, superoinferior ventricles and criss-cross atrioventricular relations.

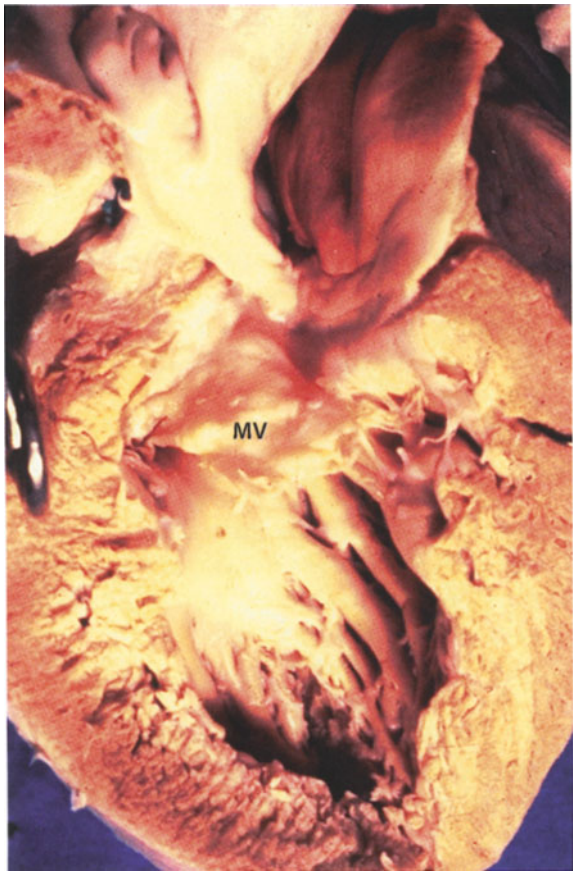

Fig. 2.13.3. Straddling mitral valve: morphology. Heart with straddling of the mitral valve (*MV* mitral valve) (photograph courtesy of Dr. Bruno Marino)

The presence of a straddling mitral valve is accompanied by severely marked malalignment of the ventricles relative to the atria, with the angle between the ventricular septum and the atrial septum in the short-axis projection averaging 150 degrees (the normal ventriculoatrial septal angle averaging 5 degrees), with hypoplasia of the right ventricular sinus (=inflow tract) with tricuspid valve stenosis or hypoplasia.

Associated anomalies

Ventricular septal defect (usually of inlet type) is present in 80% of cases, double outlet right ventricle in 30–40% of cases, complete transposition of the great arteries (25%), double discordance (=congenitally corrected transposition of the great arteries) (20%), Taussig-Bing anomaly, mitral stenosis (in 25% of cases with straddling tricuspid valve), tricuspid stenosis or hypoplasia in cases with straddling mitral valve, right (or left respectively) ventricular outflow tract obstruction, tetralogy of Fallot, superoinferior ventricles with criss-cross atrioventricular relations, dextrocardia.

Pathophysiology

Straddling of an atrioventricular valve results in maldevelopment of the ipsilateral ventricle by directing blood flow away from the ipsilateral ventricle and towards the contralateral ventricle; in the presence of severe hypoplasia of a ventricular chamber, the single ventricle physiology is the consequence. A straddling atrioventricular valve, generally competent, can also contribute to creating an obstruction to the contralateral ventricular outflow tract, because of the anomalous valve insertions.

Diagnosis

Clinical pattern: the clinical presentation depends upon the associated anomalies, and it varies from congestive heart failure to cyanosis, to a combination of the two.

Electrocardiogram: not diagnostic, is basically correlated with the ventricular morphology.

Chest X-ray: variable according to the pathophysiologic pattern.

Echocardiogram: until recently cross sectional echocardiography was the technique to provide unique information necessary for planning the rationale for the best type of surgical approach; nowadays the three-dimensional echocardiography provides diagnostic information superior to standard

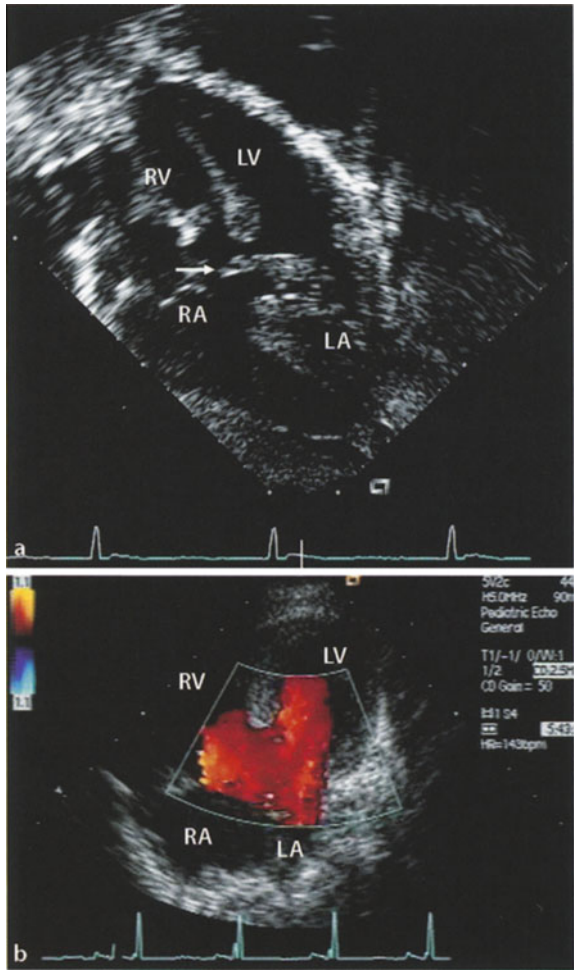

Fig. 2.13.4. Echocardiography: **a** straddling of the right atrioventricular valve (white arrow) in a patient with double outlet right ventricle, **b** Doppler echocardiography of the same patient showing the straddling of the right atrioventricular valve (white arrow) in a patient with double outlet right ventricle (*LA* left atrium, *LV* left ventricle, *RA* right atrium, *RV* right ventricle)

cross sectional techniques, particularly because it allows for exact measurement of the volumes of the respective ventricles; the short-axis subcostal view is the best projection to evaluate presence and degree of straddling of the atrioventricular valves (Fig. 2.13.4).

Cardiac catheterization: is indicated to precisely define the associated cardiac anomalies and to provide information on the hemodynamic pattern.

Indications for surgical treatment

Several surgical techniques have been utilized to perform a biventricular repair in the presence of a straddling atrioventricular valve, all aiming at preserving a normal valve functioning, including:

- baffle or slit of the prosthetic patch used for closure of the ventricular septal defect allowing the maintenance of the straddling atrioventricular valve in its normal position,
- retraction of the anomalous chordae in the ipsilateral ventricle, with attachment of the ventricular septal defect patch to the retracted tension apparatus,
- detachment of the anomalous valve insertion and reattachment to the ipsilateral side of the patch used to close the ventricular septal defect.

The presence of a straddling tricuspid valve can complicate the conventional intraventricular repair or can preclude it, therefore representing a potential contraindication to the biventricular repair, particularly in the presence of hypoplasia of the right ventricle with ventricular septal defect and one of the following:

- atrioventricular discordance,
- atrioventricular septal defect with left ventricular outflow tract obstruction,
- ventriculoarterial discordance (either transposition of the great arteries or double outlet right ventricle),
- pulmonary stenosis or atresia.

Of course the above difficult situations can be managed with increasing experience and expertise, allowing for a conventional biventricular repair.

One and half ventricular repair: in patients with reduced size of the morphologically right ventricle, a one-and-half ventricular repair is performed, with end-to-side anastomosis of the superior vena cava to the right pulmonary artery (=bidirectional Glenn) in addition to the intracardiac repair, in order

to reduce the volume overload of the hypoplastic right ventricle, inadequate to sustain the entire pulmonary circulation.

Univentricular repair: in patients with severe straddling tricuspid valve in the presence of hypoplasia of the morphologically right ventricle, atrioventricular or ventriculoarterial discordance and pulmonary stenosis or atresia, a univentricular type of repair (total cavopulmonary connection = modified Fontan procedure; see chapter "Single ventricle") should be considered, balancing the high short and intermediate term risks of a complex biventricular repair with the potential long term disadvantages of a single ventricle approach.

The presence of straddling tricuspid valve with curtain-like type of morphology is also considered an indication for univentricular type of repair, because of the very poor results obtained with biventricular repair. In addition, the presence of straddling of the mitral valve, because of the frequent association of hypoplasia of the left ventricle, particularly with ventriculoarterial discordance, is considered as an indication for univentricular repair.

Surgical treatment (on cardiopulmonary bypass)

When closure of a ventricular septal defect is required as part of a biventricular type of repair, either with ventriculoarterial concordance (isolated ventricular septal defect) as well as with ventriculoarterial discordance (double outlet right ventricle or transposition of the great arteries), the presence of straddling of the tricuspid valve demand special surgical techniques to close the defect without interfering with the valve function.

In isolated tricuspid valve straddling of types A and B, the prosthetic (PTFE, Dacron, Teflon) patch used to close the ventricular septal defect is adjusted on the right ventricular side above the straddled chordae and/or papillary muscle. In isolated tricuspid valve straddling of type C, the prosthetic patch is sewn over the papillary muscle by applying it on the interventricular septum.

In the presence of double (mitral and tricuspid) straddling, the interventricular septum is incised between the two papillary muscles, and an ellipsoid-shape prosthetic patch is utilized to close the ventricular septal defect, rerouting each subvalvular apparatus into its respective ventricular chamber.

In the presence of abnormal insertion of chordae in the left ventricular outflow tract inserted on the aortic conus (or pulmonary conus with associated ventriculoarterial discordance), the conus can be incised and tailored to create a flap, obtaining an unobstructed left ventricular outflow tract.

When required, the subvalvular apparatus can be resected and then reattached to the prosthetic patch used to close the ventricular septal defect.

Potential complications

Residual or recurrent atrioventricular valve dysfunction can follow any attempt at biventricular type of repair with patch closure of the ventricular septal defect and surgical handling of the straddling valve. Residual or recurrent ventricular septal defect, arrhythmias, complete atrioventricular block, right or left ventricular outflow tract obstruction are possible after biventricular repair.

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Chapter 2.14 Isomerism

Incidence

The incidence is reported between 0.1 and 0.4% of congenital heart defects. A tendency for right isomerism to affect males, and for left isomerism to affect females has been reported.

Morphology

In patients with incomplete lateralization of thoracic and visceral organs, the atrial appendages are also not lateralized, and in the vast majority of these patients both atrial appendages present with similar internal and external morphology, characteristic of either the right or the left atrial appendage. Atrial isomerism is the designation for these hearts with bilaterally right atrial appendages (Fig. 2.14.1) or bilaterally left atrial appendages (Fig. 2.14.2).

Anomalous systemic venous connections are generally associated with atrial isomerism, and understanding isomerism is important in sorting out the various lesions involved.

These so-called heterotaxic syndromes are characterized by failure of many "right-left" differentiations, leading to ambiguity in the visceroatrial situs, along with anomalies of systemic and/or pulmonary venous connections.

In patients with *left atrial isomerism* the infra-hepatic portion of the inferior vena cava is frequently (75% of the cases) absent (=inferior vena cava interruption), and the venous return from the lower part of the body reaches the superior vena cava via the azygos vein (=azygos continuation) or via the hemiazygos vein (=hemiazygos continuation) emptying into either a right-sided superior vena cava or into a persistent left superior vena cava. Inferior vena cava interruption has never been observed in *right atrial isomerism*.

In patients with *right atrial isomerism* the right and left hepatic veins may enter the ipsilateral sides of the common atrium, remaining separate from the connection of the inferior vena cava.

Persistent left superior vena cava is present in 50% of patients with *right atrial isomerism* and in 70% of patients with *left atrial isomerism*, and in both situations, particularly in *right atrial isomerism*, it can be connected to the upper left side of the left atrium instead of the coronary sinus.

The coronary sinus orifice can often be absent, more frequently in *right atrial isomerism* than in *left atrial isomerism*.

Abnormalities of the pulmonary veins are also common in both left and right atrial isomerism; total anomalous pulmonary venous connection to the superior or inferior vena cava is more frequent in *right atrial isomerism* (in 40% of these patients with obstruction to the pulmonary venous return), whereas anomalous pulmonary venous connection into the same side of the atrium as the systemic venous drainage is more frequent in *left atrial isomerism* (generally unobstructed).

The atrial septum and ventricular septum are very rarely normal in patients with atrial isomerism; common atrium is present in about 50% of the cases, atrioventricular sep-

A. F. Corno, *Congenital Heart Defects*

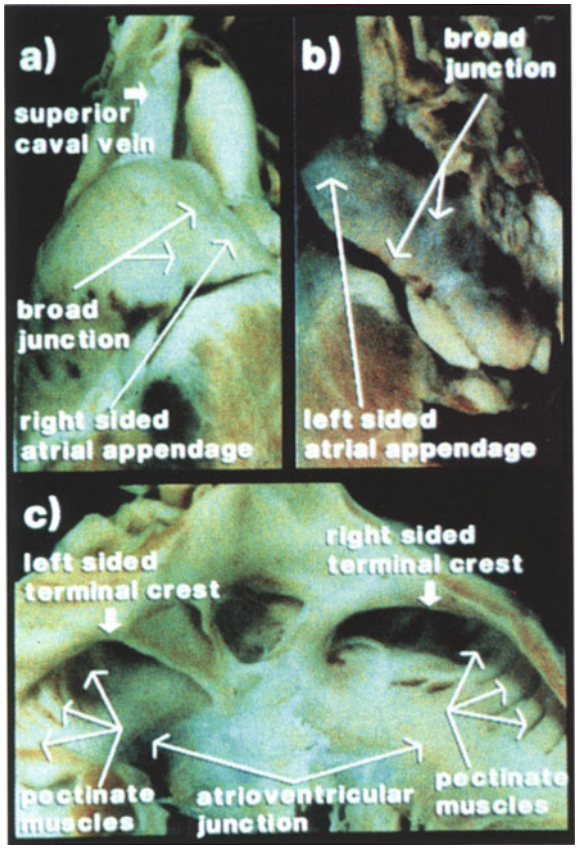

Fig. 2.14.1. Atrial isomerism: morphology. **a**, **b** External view, with both auricular appendages are blunt and both join in the atrial chamber along a broad front, **c** internal view of the atrial chambers, showing both atria with a well-formed terminal crest; the pectinate muscles encircle the atrioventricular junction, occupying the entire lateral wall of the right and left sided chambers (reproduced with permission from Rossi G, Corno AF, Montemurro G (1992) Prenatal diagnosis of isomerism of the right atrial appendages. Cardiol Young 2:298–301)

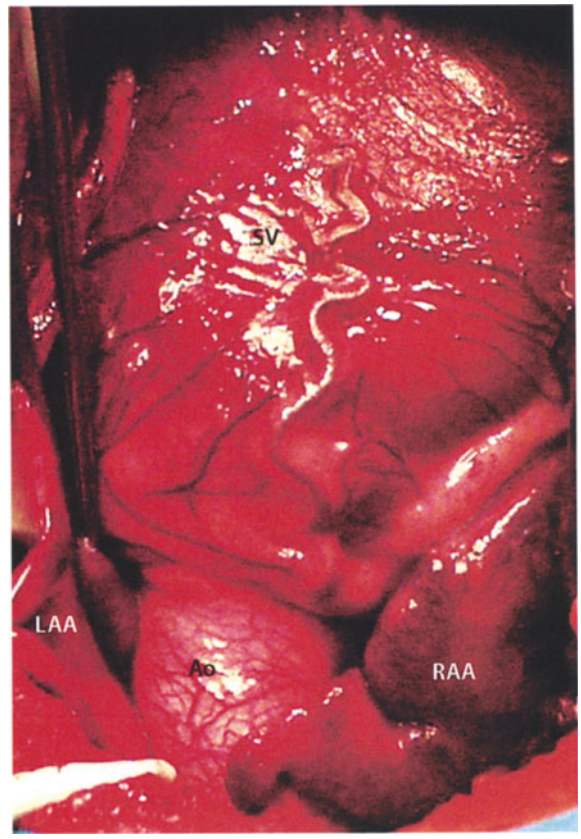

Fig. 2.14.2. Atrial isomerism: morphology. Intraoperative photograph in a patient with left atrial isomerism, interruption of the inferior vena cava with azygos continuation, two morphological left auricular appendages, common atrioventricular valve, single ventricle, anterior aorta, pulmonary stenosis. The auricular appendage on the right side (RAA) has an evident morphology of left type, as well as the one on the left side, shown with the forceps (*Ao* aorta, *LAA* left auricular appendage, *RAA* right auricular appendage, *SV* single ventricle)

tal defect in about 80% of cases, with most patients having a common atrioventricular orifice, and various types of ventricular septal defect can be present, in the vast majority of cases of atrioventricular type.

Frequently there is outflow obstruction to pulmonary arterial blood flow at the valvular and/or subvalvular level. Pulmonary atresia is slightly more common with *right atrial isomerism*, whereas pulmonary stenosis is more common in *left atrial isomerism*.

Ventriculoarterial discordance is very frequent (75–90% of cases), in one third of patients with double outlet right ventricle.

Pulmonary artery anomalies are not rare, particularly when there is pulmonary atresia with the ductus arteriosus as the only source of pulmonary blood flow. After closure of the ductus arteriosus, a "coarctation" commonly develops at the origin of the left pulmonary artery, just at the insertion of the ductus arteriosus.

The branching pattern of the pulmonary arteries generally assumes one of two forms, depending on whether left or right atrial isomerism is present. In *right atrial isomerism* both right and left pulmonary arteries tend to look like a normal right pulmonary artery

(=two right pulmonary arteries), with the bronchus for the upper lobe being above the first segmental artery for the right upper lobe (epiarterial bronchus). In contrast, in *left atrial isomerism* the bronchus is below the pulmonary artery at the hilum (hypoarterial bronchus), as is the case for a normal left pulmonary artery (=two left pulmonary arteries).

Right atrial isomerism is generally characterized by the presence of bilateral sinus nodes, one in each atrium; two atrioventricular nodes may also be present. In *left atrial isomerism* there is absence of sinus node in the majority of patients, while less frequently the sinus node is in an anomalous position and usually hypoplasic. Atrial isomerism generally corresponds to thoracic isomerism; therefore, in *right atrial isomerism* both lungs tend to be trilobed (=two right lungs), whereas in *left atrial isomerism* both lungs tend to be bilobed (=two left lungs). Finally, *asplenia* is more commonly present in *right atrial isomerism*, whereas *polysplenia* is more frequently associated with *left atrial isomerism*. These features have contributed to the general rule (with several exceptions) that patients with *right atrial isomerism* tend to have bilateral "right-sidedness" (asplenia), whereas those with *left atrial isomerism* tend to have bilateral "left-sidedness" (polysplenia).

Because of the extreme morphological variability within the cases with atrial isomerism, the term "heterotaxy" has been suggested to define the presence of any of the numerous possible anomalies of lateralization.

Associated anomalies

Left isomerism: polysplenia, anomalous systemic and/or pulmonary venous connections are very frequent, with interruption of the inferior vena cava and azygos continuation as the most frequent (56–92% of cases), followed by anomalous pulmonary venous connection (56%) that in a certain percentage of patients is potentially obstructive, common atrioventricular valve (46–49%), common atrium (38%), cor triatriatum (30%), pulmonary atresia or stenosis (28%), aortic coarctation (16%), congenital atrioventricular block (7%). Polysplenia is frequently accompanied by extrahepatic biliary atresia.

Right isomerism: asplenia, valvular and subvalvular pulmonary stenosis or pulmonary atresia are predominant (89% of cases), followed by discordant ventriculoarterial connection (72–75%) either because of the presence of transposition of the great arteries or double outlet right ventricle, atrioventricular septal defect (72%) with or without a common atrium, single ventricle (55%), extracardiac total anomalous pulmonary venous connection (50%), persistent left superior vena cava, bilateral right auricular appendages (20%). Asplenia contributes to render the patient more susceptible to infections.

Pathophysiology

The pathophysiological pattern totally depends upon the combination of intracardiac defects, particularly by the presence and degree of reduction of the pulmonary blood flow, right-to-left intracardiac shunt, atrioventricular valve regurgitation, and obstruction to the systemic outflow.

The impact of asplenia goes beyond the associated congenital heart defects, with the obvious implications related to blood infection and sepsis.

Diagnosis

Clinical pattern: cyanosis is present in the vast majority (96%) of patients, generally from the first month of life; congestive heart failure with or without cyanosis is also very frequent; cell blood count reveals the presence of Howell-Jolly bodies, inclusions in the erythrocytes, in the presence of asplenia.

Electrocardiogram: abnormal P wave axis, left-ward and superior, is present in 50–75% of patients with left atrial isomerism; negative P waves in leads II, III, and aVF (so-called coronary sinus rhythm); abnormalities of the conduction system, including complete atrioventricular block (10% of cases with left atrial isomerism), sick sinus syndrome and supraventricular tachycardia, are more frequent in left than in right isomerism.

Chest X-ray: anteroposterior view determines the thoracic situs, the cardiac size and location (to show dextrocardia), the bronchial anatomy (to show left or right bronchi) and the abdominal status (to show situs viscerum inversus); lateral view allows definition of right isomerism, when each pulmonary artery is anterior to the respective bronchus, or left isomerism, when each pulmonary artery is superior and posterior to the respective bronchus; typical is the appearance of a symmetrical liver on the chest and abdominal X-ray, and a discordant position between the cardiac apex and gastric air bubble; the absence of the inferior vena cava shadow on the lateral projection may suggest azygos continuation of the inferior vena cava.

Echocardiogram: echocardiography can show the anomalous positional and spatial relationship between the inferior vena cava and the abdominal aorta (Fig. 2.14.3), both on the same side of the spine in patients with asplenia (Fig. 2.14.4); a different relationship exists in patients with polysplenia (Fig. 2.14.5); at the level of the diaphragm a mirror image of the normal situation (inferior vena cava to the right and slightly anterior to the descending aorta) exists in patients with situs inversus (Fig. 2.14.6); direct imaging of the morphology of the atrial appendages is feasible, as well as of the mode of connection of the hepatic veins to the atria; absence of intrahepatic portion of the inferior vena cava is associated with azygos continuation in left isomerism; once pres-

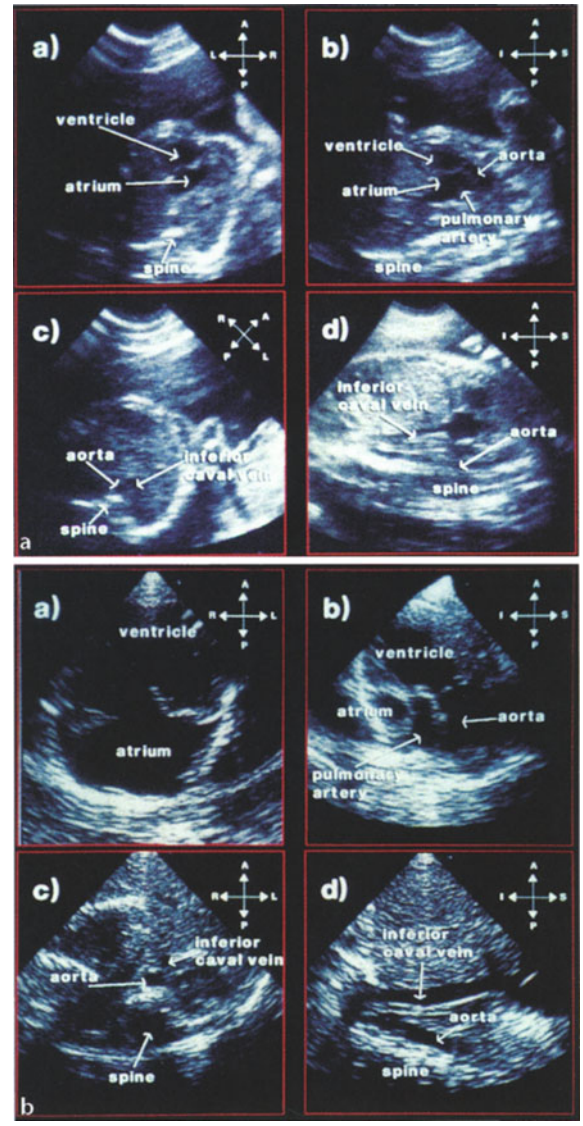

Fig. 2.14.3. Atrial isomerism: echocardiography. **a** Prenatal echocardiography at 27 weeks of gestation, showing a common atrium connected through a common atrioventricular valve to a ventricle of right morphology (**a**), giving origin to both the great arteries with the aorta anterior to the pulmonary artery and subpulmonary obstruction (**b**); the short-axis view shows that the inferior vena cava and the abdominal aorta are on the same side of the spine, with the vein anterior and to the left of the artery (**c** and **d**), arrangement consistent with right isomerism, **b** neonatal echocardiography of the same subject as in **a**, with short-axis view confirming the prenatal echocardiographic diagnosis (reproduced with permission from: Rossi G, Corno AF, Montemurro G (1992) Prenatal diagnosis of isomerism of the right atrial appendages. Cardiol Young 2:298–301)

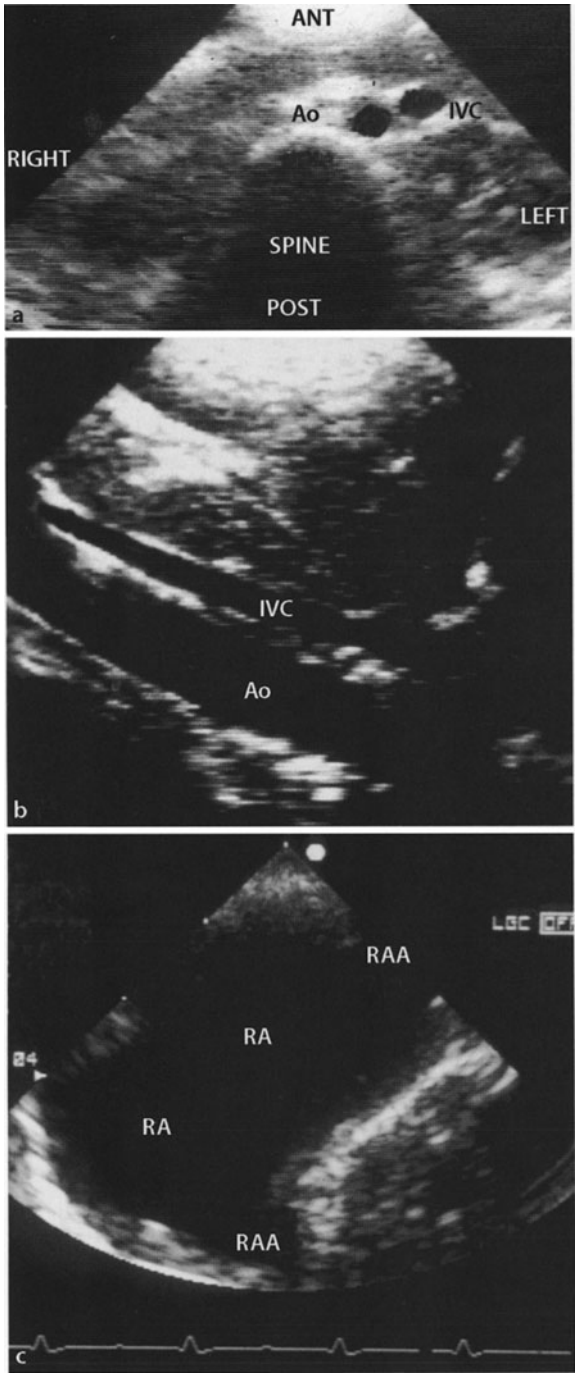

Fig. 2.14.4. Atrial isomerism: echocardiography. **a** Subcostal horizontal section showing the relationship between spine with aorta and inferior vena cava in a neonate with *right* isomerism (*Ao* aorta, *IVC* inferior vena cava), **b** subcostal sagittal section showing the relationship between aorta and inferior vena cava in a neonate with *right* isomerism, **c** transesophageal echocardiography showing the *right* atrial isomerism in a neonate with double inlet right ventricle (*RA* right atrium, *RAA* right auricular appendage) (photographs courtesy of Dr. Michael Rigby)

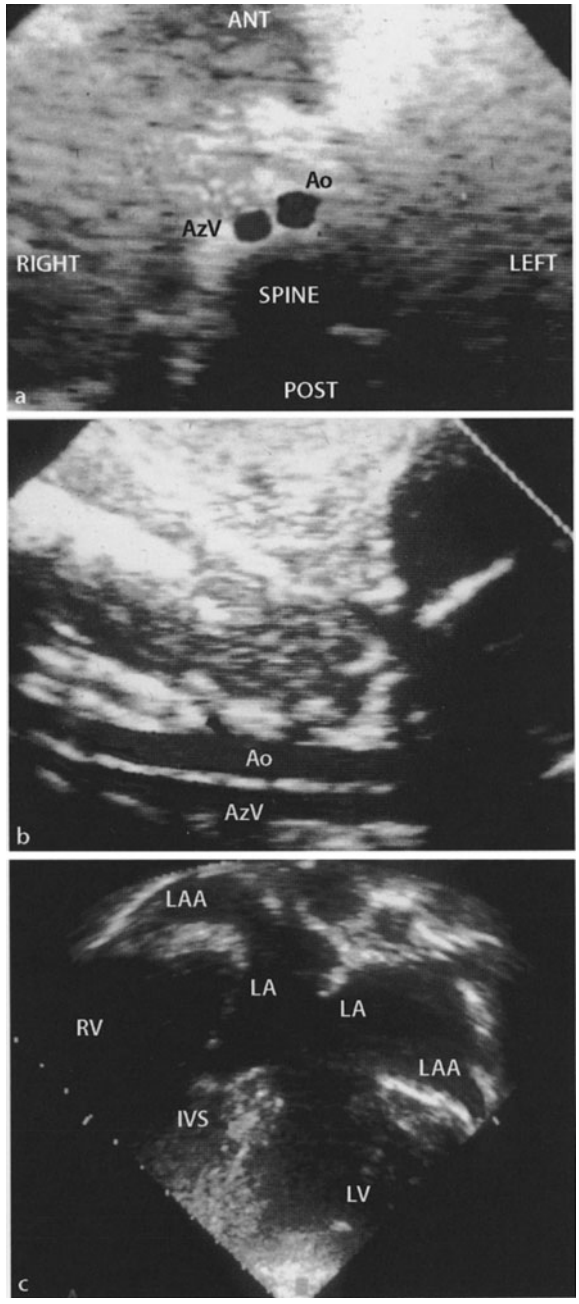

Fig. 2.14.5. Atrial isomerism: Echocardiography. **a** Subcostal horizontal section showing the relationship between the spine with an aorta and inferior vena cava in a neonate with *left* isomerism, interruption of the inferior vena cava and azygos continuation (*Ao* aorta, *AzV* azygos vein), **b** subcostal sagittal section showing the relationship between the aorta and inferior vena cava in a neonate with *left* isomerism, interruption of the inferior vena cava and azygos continuation, **c** showing the atrial arrangement in a neonate with *left* atrial isomerism and complete atrioventricular septal defect (*IVS* interventricular septum, *LA* left atrium, *LAA* left auricular appendage, *LV* left ventricle, *RV* right ventricle) (photographs courtesy of Dr. Michael Rigby)

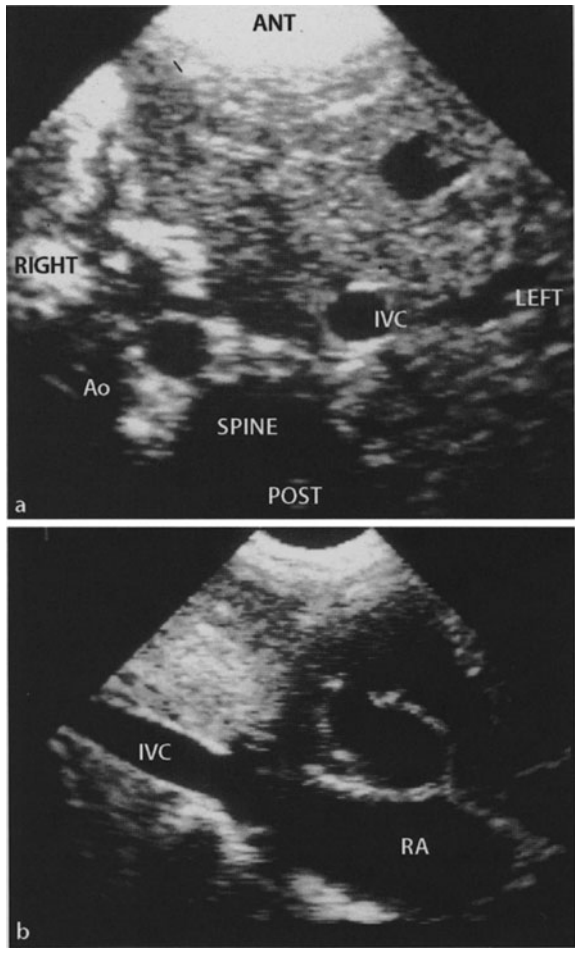

Fig. 2.14.6. Atrial isomerism: echocardiography. **a** Subcostal horizontal section showing the relationship between the spine with an aorta and inferior vena cava in a neonate with *situs inversus*, **b** subcostal sagittal section showing the left-sided position of the inferior vena cava in a neonate with *situs inversus* (*IVC* inferior vena cava, *RA* right atrium) (photographs courtesy of Dr. Michael Rigby)

ence and type of isomerism have been confirmed, echocardiography will show the associated intracardiac defects.

Cardiac catheterization: particularly important to study the systemic and pulmonary venous connections (Figs. 2.14.7–2.14.10), when this information is not provided in a reliable way by echocardiography; useful also to rule out obstructions to the pulmonary venous return and increased pulmonary vascular resistance.

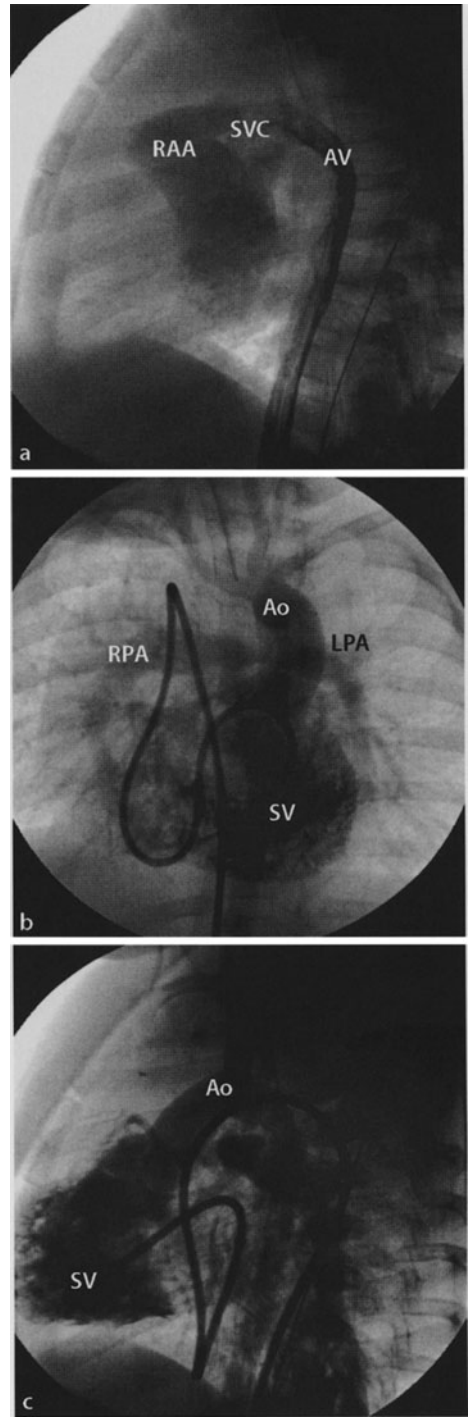

Fig. 2.14.7. Atrial isomerism: angiography. **a** Lateral view injection in the azygos continuation, with opacification of the superior vena cava and of the right auricular appendage, with evident morphology of the left type (*AV* azygos vein, *RAA* right auricular appendage, *SVC* superior vena cava), **b** anteroposterior view: injection in the single ventricle, with opacification of the anterior aorta and of the two pulmonary arteries, both with morphology of the left type, and stenotic

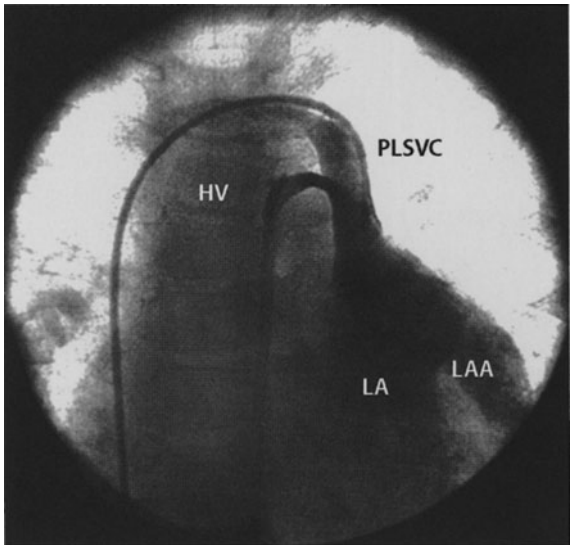

Fig. 2.14.8. Atrial isomerism: angiography. Anteroposterior view with contrast injection showing the persistent left superior vena cava draining into the left atrium (*HV* hemiazygos vein, *LA* left atrium, *LAA* left auricular appendage, *PLSVC* persistent left superior vena cava)

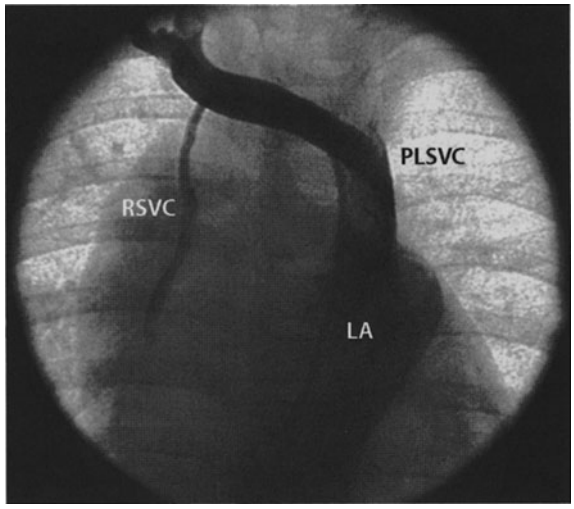

Fig. 2.14.9. Atrial isomerism: angiography. Anteroposterior view with contrast injection in the innominate vein showing a very hypoplastic right superior vena cava and a persistent left superior vena cava draining into a left-sided morphological right atrium (*HV* hemiazygos vein, *LA* left atrium, *PLSVC* persistent left superior vena cava, *RSVC* right superior vena cava)

Indications for surgical treatment

Without treatment the congenital heart defects associated with isomerism are accompanied by a mortality between 50 and 95% in the first year of life, according to the relevant hemodynamic pattern. Surgical treatment has improved the survival rate, particularly during infancy, but the 5 year mortality remains as high as 30 to 50%. The lesions associated with left isomerism can often be successfully corrected, with a combination of intraatrial and intraventricular rerouting, with a biological valved conduit to reconstruct the continuity between the right ventricle and the pulmonary artery. The lesions associated with right isomerism generally preclude biventricular repair, require a staged palliation and carry a poorer prognosis.

Historically the procedures of total cavopulmonary connection in patients with heterotaxy syndrome, right atrial isomerism and functionally univentricular heart have been associated with high mortality and morbidity. Nowadays, with the improved knowledge of the adequate timing for the staged surgical approach for univentricular hearts, and with the improved perioperative management, the results in this complex group of patients have also substantially improved. The staged approach can require either palliative surgical procedures to increase the pulmonary blood flow (modified Blalock-Taussig shunt) or to decrease pulmonary blood flow and pressure (pulmonary artery banding) in order to allow for subsequent univentricular type of repair (see chapter "Single ventricle").

confluence; the catheter reaches the ventricular cavity from the azygos vein, superior vena cava, right atrium (*Ao* aorta, *RPA* right pulmonary artery, *LPA* left pulmonary artery, *SV* single ventricle), **c** lateral view: injection in the single ventricle, with opacification of the anterior aorta and of the two pulmonary arteries; the catheter reaches the ventricular cavity from the azygos vein, superior vena cava, right atrium

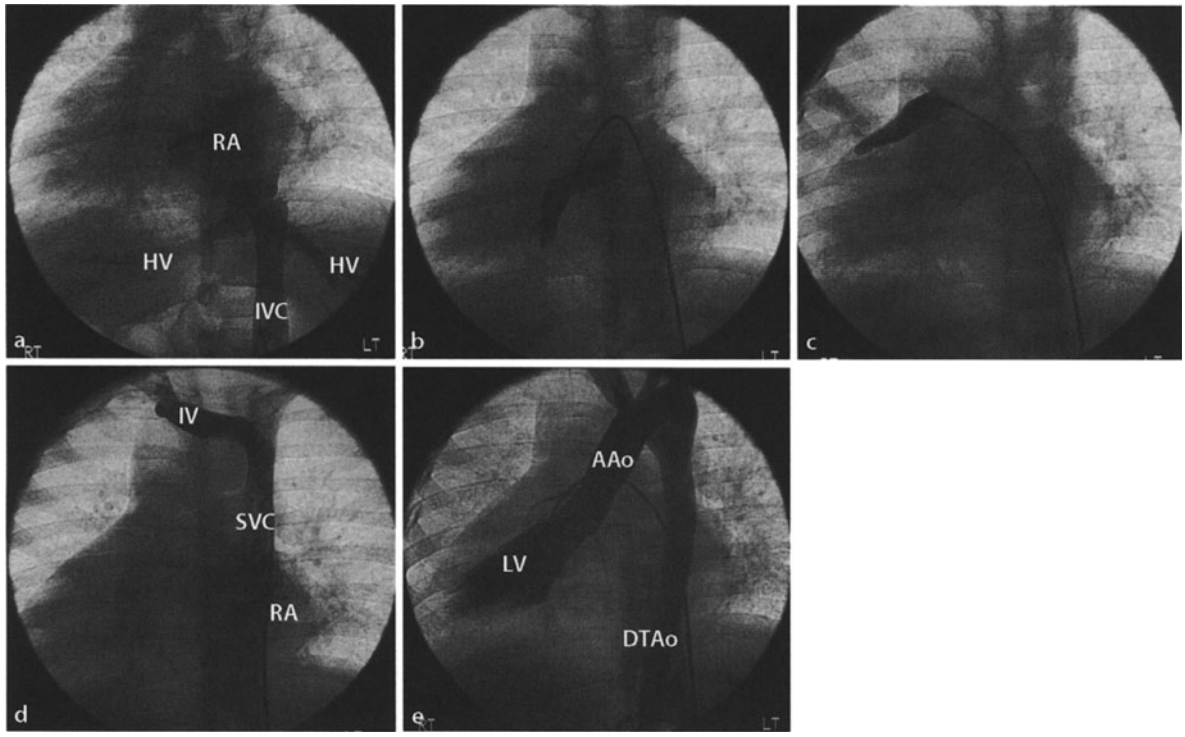

Fig. 2.14.10. Atrial isomerism: angiography. Anteroposterior view with contrast injection in **a** the inferior vena cava located on the left side of the spine, with opacification of the hepatic veins and the right atrium, located on the left side, in a child with situs viscerum inversus (*HV* hepatic veins, *IVC* inferior vena cava, *RA* right atrium), **b** the right auricular appendage of left type morphology and located on the right side of the spine, **c** the left auricular appendage of left type morphology and located on the right side of the spine, **d** the innominate vein, showing the superior vena cava and the right atrium located on the left side, in the same child as in **a**, **b** and **c** with situs viscerum inversus and left atrial isomerism (*SVC* superior vena cava, *IV* innominate vein), **e** the left ventricle located on the right side of the spine, with opacification of the aorta confirming the diagnosis of situs viscerum inversus (*AAo* ascending aorta, *DTAo* descending thoracic aorta, *LV* left ventricle)

Surgical treatment

Palliation:

- Pulmonary artery banding: see chapter "Ventricular septal defect".
- Modified Blalock-Taussig shunt: see chapter "Tetralogy of Fallot".

Repair: for the operations performed on cardiopulmonary bypass, to avoid deep hypothermia and circulatory arrest (therefore a single venous cannula to drain the venous return), an individualized technique of venous cannulation, adapted to the anomalies of the systemic venous return of each patient, is required.

Biventricular repair: in the presence of anomalous systemic and/or pulmonary venous connection(s) a complex interatrial baffle (pericardium, Teflon, PTFE) very frequently is required to obtain adequate rerouting of the systemic and pulmonary venous returns (Fig. 2.14.12). In the presence of persistent left superior vena cava, particularly when it is connected to the left atrium (Fig. 2.14.8), the extracardiac connection of the left superior vena cava either to the right superior vena cava or to the right auricular appendage can simplify the partitioning of the atrial chambers. The technique to close atrioventricular or ventricular septal defect have been described in the respective chapters.

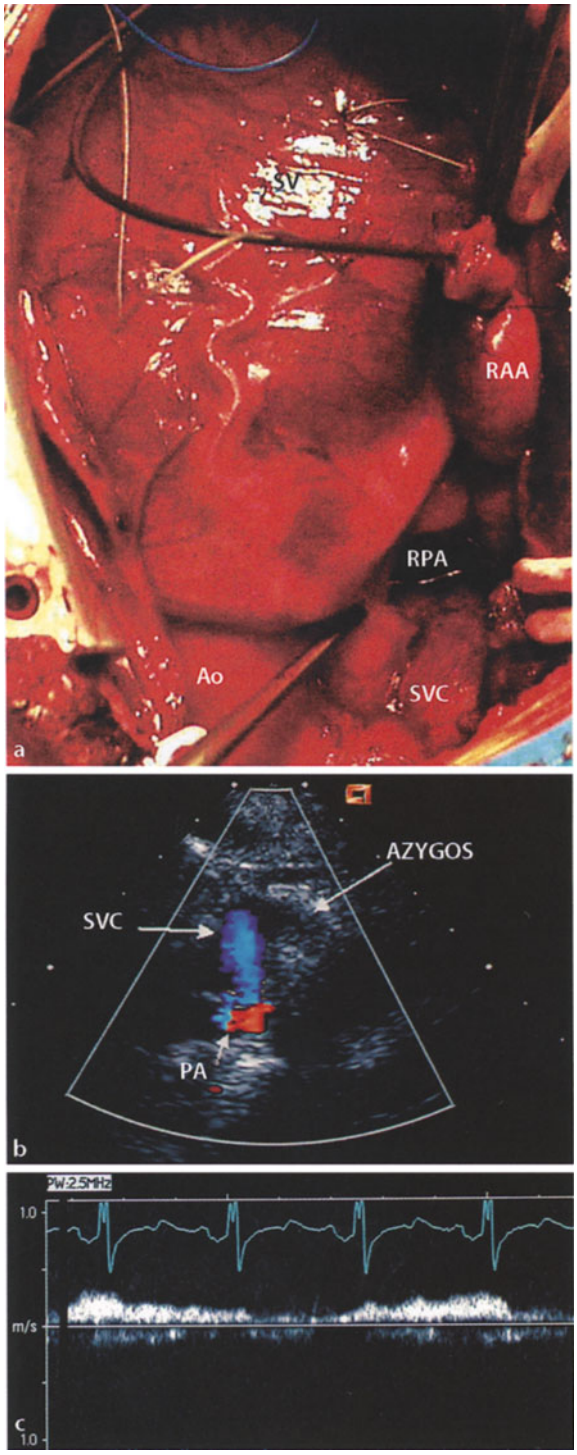

Fig. 2.14.11. Atrial isomerism: surgery. **a** Intraoperative photograph (same patient as in Fig. 2.14.2) at the end of a Kawashima operation (= end-to-side anastomosis of the superior vena cava to the right pulmonary artery, without transection of the azygos vein) (*Ao* aorta, *RAA* right auricular appendage, *RPA* right pulmonary artery, *SV* single ventricle, *SVC* superior vena cava), **b** postoperative echocardiography (same patient as in Fig. 2.14.2) after the Kawashima operation, showing both the azygos vein and the superior vena cava connected to the pulmonary artery (*AZYGOS* azygos vein, *PA* pulmonary artery), **c** postoperative Doppler echocardiography (same patient as in Fig. 2.14.2) after the Kawashima operation, showing the continuous flow in the azygos vein (photographs **b** and **c** courtesy of Dr. Nicole Sekarski)

Univentricular repair:

- Bidirectional Glenn or Hemi-Fontan: see chapter "Single ventricle".
- Modified Fontan or total cavopulmonary connection: see chapter "Single ventricle".
- In the presence of interruption of the inferior vena cava and azygos continuation, the preferred surgical option is the *Kawashima operation* (Fig. 2.14.11 a), consisting in the end-to-side anastomosis between the transected superior vena cava and the right pulmonary artery, like in the bidirectional Glenn, but without dividing the azygos vein (Fig. 2.14.11 b and c); in this way the entire systemic venous return, excluding splanchnic veins and coronary sinus, is deviated directly into the pulmonary circulation. The Kawashima operation therefore represents an almost complete univentricular type of repair, like the total cavopulmonary connection, see page 188.

Potential complications

The main differences in comparison with the patients with functionally univentricular hearts without isomerism with regard to the complications are the following:

- after bidirectional Glenn (see chapter "Single ventricle"), a higher incidence of prolonged pleural effusions and chylothorax,
- after modified Fontan or total cavopulmonary connection (see chapter "Single ventricle"), a higher incidence of prolonged pleural effusions, supraventricular arrhythmias, pulmonary arteriovenous malformations and venovenous collaterals (particularly after *Kawashima operation*).

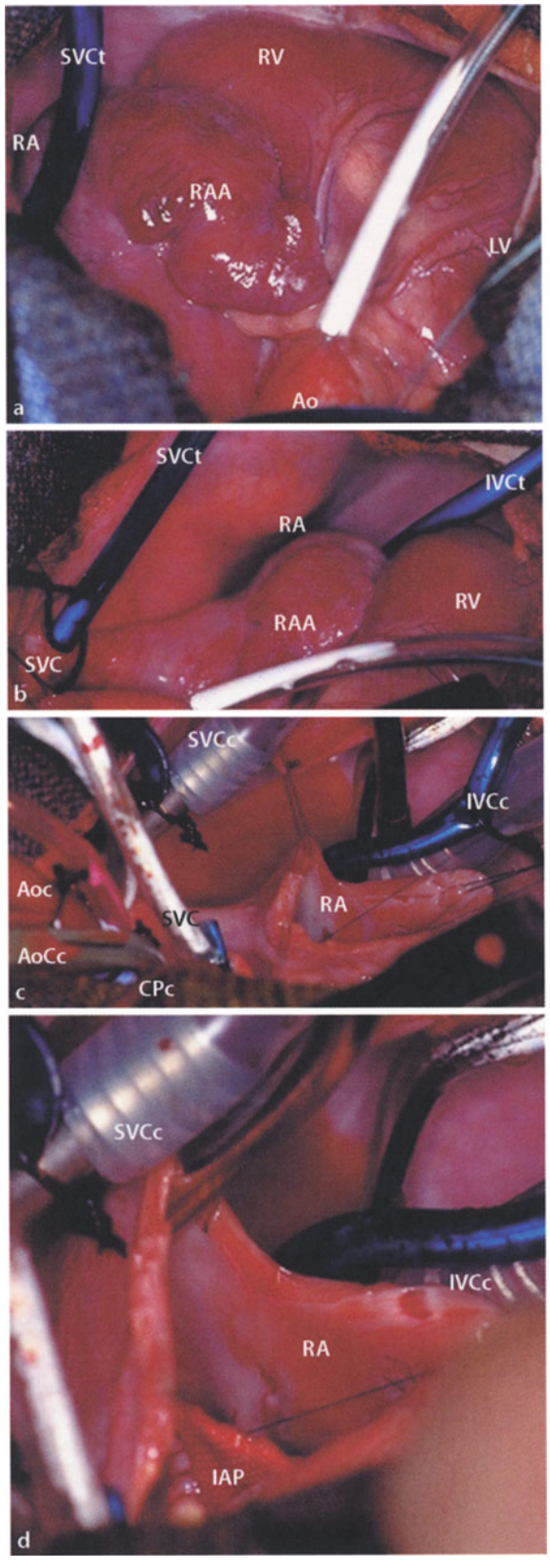

Fig. 2.14.12. Atrial isomerism: surgery. Intraoperative photograph (same child as in Fig. 2.14.10) taken from **a** the head of the patient showing the situs inversus of the heart with dextrocardia, the superior vena cava and the right atrium located on the left side, and the left morphology of the right auricular appendage, located on the left side, because of the left atrial isomerism (*Ao* aorta, *LV* left ventricle, *RA* right atrium, *RAA* right auricular appendage, *RV* right ventricle, *SVCt* superior vena cava tourniquet), **b** the right side with a different view of the same patient showing the superior vena cava and the right atrium located on the left side (*IVCt* inferior vena cava tourniquet, *SVC* superior vena cava), **c** the right side showing the cannulation of aorta, left sided superior and inferior vena cava, with right atriotomy for intracardiac repair (*Aoc* aortic cannula, *AoCc* aortic cross clamp, *CPc* cardioplegia cannula, *IVCc* inferior vena cava cannula), **d** the right side showing the prosthetic patch used for interatrial partitioning (*IAP* interatrial patch)

In patients with isomerism undergoing a total cavopulmonary connection a larger use of fenestration could probably prevent or reduce this high incidence of postoperative complications and further improve the outcome.

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Chapter 2.15 Slings and rings

In this chapter the two most frequent anatomical situations with slings and/or rings are taken into considerations: pulmonary artery sling and vascular ring.

Pulmonary artery sling

Incidence

Airway stenosis is present in 1.2% of children with congenital heart defects, and is more frequent in patients with Down syndrome (40%) than in the normal population. No gender prevalence has been detected. The incidence of the pulmonary artery sling has not been precisely quantified, although it is very rare.

Morphology

Pulmonary artery sling (=anomalous left pulmonary artery) is a rare congenital vascular anomaly in which the left pulmonary artery originates extrapericardially from the posterior aspect of the right pulmonary artery, encircles the right main bronchus and then courses from right to left, posterior to the distal trachea and tracheal bifurcation and anterior to the esophagus, before entering the hilum of the left lung (Fig. 2.15.1).

The left pulmonary artery is often relatively hypoplastic and considerably smaller than the right pulmonary artery, which appears larger than normal, virtually a direct extension of the main pulmonary artery.

The ligamentum arteriosum passes posteriorly to the aorta from the point of origin of the right pulmonary artery from the main pulmonary artery, superiorly to the left main bronchus, effectively creating a vascular ring (or sling) around the trachea but not around the esophagus.

Associated anomalies

Approximately 50% of patients with a pulmonary artery sling have complete cartilaginous tracheal rings: the posterior membranous component of the trachea is absent, and the tracheal cartilages, rather than being U shaped, are O shaped. The presence of complete tracheal rings does not imply that important stenosis will necessarily be present, although the trachea is often narrower than normal. The complete rings may be localized to the region where the sling passes around the trachea, although often they extend for the entire length of the trachea, creating a long-segment tracheal stenosis. In the area where the sling passes around the trachea, there is likely to be tracheal compression resulting in important functional stenosis, even if there is not an underlying anatomic stenosis.

Bronchus suis (="pig bronchus"), consisting of separate high origin of the epiarterial bronchus to the right upper lobe from the trachea, is also a relatively frequent association of pulmonary artery sling.

Congenital heart defects are present in 50% of patients with pulmonary artery sling, most commonly atrial septal defect, ventricular

A. F. Corno, *Congenital Heart Defects*

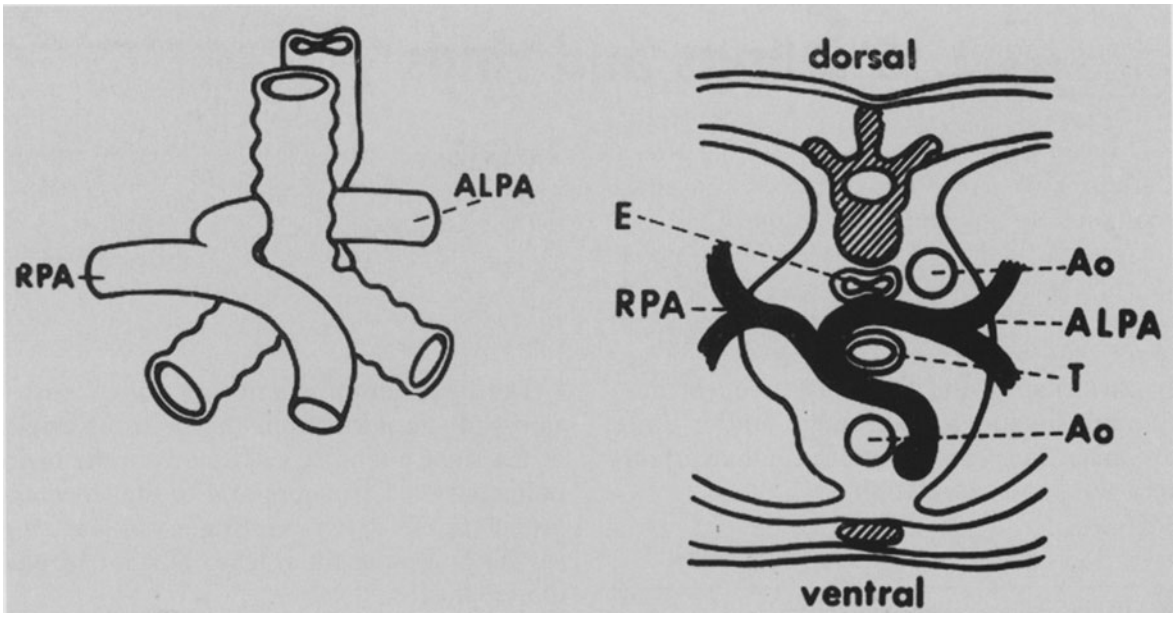

Fig. 2.15.1. Pulmonary artery sling: morphology. Schematic drawing of the pulmonary artery sling and the relationship between the anomalous left pulmonary artery and the surrounding structures (*ALPA* anomalous left pulmonary artery, *Ao* aorta, *E* esophagus, *RPA* right pulonay artery, *T* trachea)

septal defect, patent ductus arteriosus, persistent left superior vena cava, scimitar syndrome (see chapter "Partial anomalous pulmonary venous connection"). Extremely rare is the association with tricuspid atresia, single ventricle, tetralogy of Fallot, transposition of the great arteries, aortic arch anomalies.

Other anomalies in the arterial supply to one or both lungs can be seen in association with this malformation, like the anomalous left pulmonary artery only supplying the left upper lobe with the normal pulmonary artery supply to the left lower lobe, or partial anomalous supplying the right upper lobe from an anomalous left pulmonary artery.

Pathophysiology

The anomalous left pulmonary artery causes compression of the right main bronchus and distal trachea, with deviation of the distal trachea to the left and subsequent compression of the right main bronchus and distal trachea. The resulting airway obstruction affects primarily the right lung, although compression of the lower trachea and left main bronchus can result in bilateral obstruction. When there is a prolonged duration of the airway compression, tracheomalacia and/or bronchomalacia can be a severe consequence.

Diagnosis

Clinical pattern: clinical signs generally (90% of cases) present within the first month of life with upper airway symptoms, obstructive emphysema, lung atelectasis, severe respiratory distress, expiratory stridor, wheezing, recurrent respiratory infections, obstructive apnea, tachypnea, cyanosis; all these symptoms are mostly episodic and variable; symptoms of esophageal compression (dysphagia) are rarely present.

Electrocardiogram: right ventricular hypertrophy can be present.

Chest X-ray: deviation of the lower trachea to the left; hyperinflation of the right lung, sometimes accompanied by hyperinflation

also of the left lung; atelectasis of a lobe or of an entire lung may be present; the lateral view shows a density anterior to the esophagus and posterior to the trachea just above the carina, due to the anomalous left pulmonary artery; barium swallow is the diagnostic procedure of choice: an anterior indentation of the esophagus on the lateral projection is diagnostic of pulmonary artery sling; in the frontal projection the esophagus is displaced to the right and demonstrates an oblique impression immediately posterior to the carina.

Echocardiogram: suprasternal view shows absence of the normal bifurcation of the pulmonary arteries, with the left pulmonary artery apparently absent; examination of the right pulmonary artery reveals the left pulmonary artery arising from its posterior surface; the anomalous left pulmonary artery can be followed posterior to the trachea and anterior to the esophagus.

Cardiac catheterization: injection in the main pulmonary artery is the gold standard for precise diagnosis.

Combined bronchoesophagoscopy: it should be systematically used to evaluate the extent of the airway compromise, in order to decide if the reimplantation of the anomalous left pulmonary artery needs to be accompanied by simultaneous tracheal surgery; the endoscopy can show a stenosis with circular tracheal rings, pulsatile compression of the lateral tracheal wall, tracheobronchomalacia of various degrees, subglottic stenosis, anomalies of the bronchial distribution, pulsatile compression of the esophagus, rarely other tracheoesophageal malformations; it also allows anticipation of problems during the tracheal intubation, as well as postoperative extubation problems in the case of a nontreated associated airways obstruction.

Computed tomography and magnetic resonance imaging: define the anatomical details of both the airway and the vascular structures; in particular it demonstrates the left pulmonary artery arising from the right pulmonary artery, encircling the trachea, and reaching the hilum of the left lung coursing anteriorly to the esophagus and the aorta (Fig. 2.15.2); furthermore, it shows the presence of complete tracheal rings and the level and the extent of tracheal stenosis (Fig. 2.15.3).

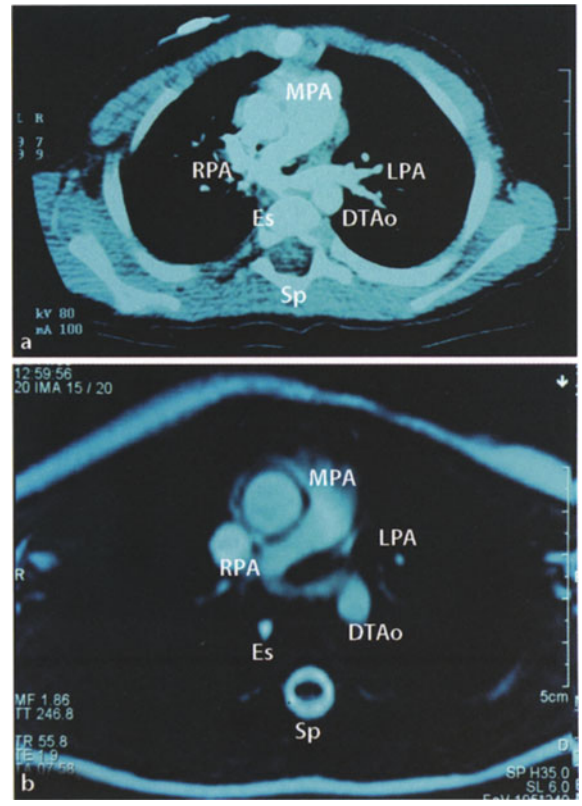

Fig. 2.15.2. Pulmonary artery sling: radiology. **a** CT scan showing the left pulmonary artery arising from the right pulmonary artery, encircling the trachea, and reaching the hilum of the left lung coursing anteriorly to the esophagus and the aorta (*DTAo* descending thoracic aorta, *Es* esophagus, *LPA* left pulmonary artery, *MPA* main pulmonary artery, *RPA* right pulmonary artery, *Sp* spine), **b** contrast magnetic resonance in the same child showing the left pulmonary artery arising from the right pulmonary artery, encircling the trachea, and reaching the hilum of the left lung coursing anteriorly to the esophagus and the aorta,

Indications for surgical treatment

Most infants with this anomaly present with severe respiratory distress within the first few months of life. Since elevated mortality

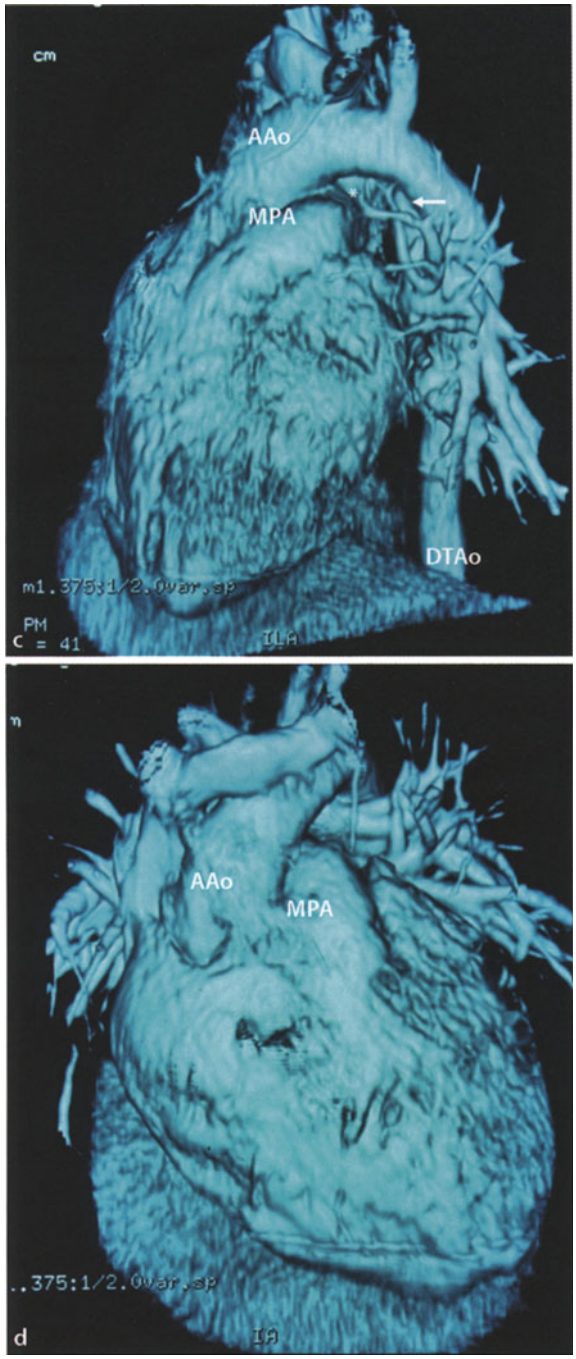

Fig. 2.15.2. **c** lateral projection of the 3D reconstruction of the CT scan in the same patient showing the absence of origin of the left pulmonary artery from the main pulmonary artery, and its origin (white arrow) from the right pulmonary artery (white star) (*AAo* ascending aorta, *DTAo* descending thoracic aorta), **d** anteroposterior projection of the 3D reconstruction of the CT scan showing the absence of origin of the left pulmonary artery from the main pulmonary artery

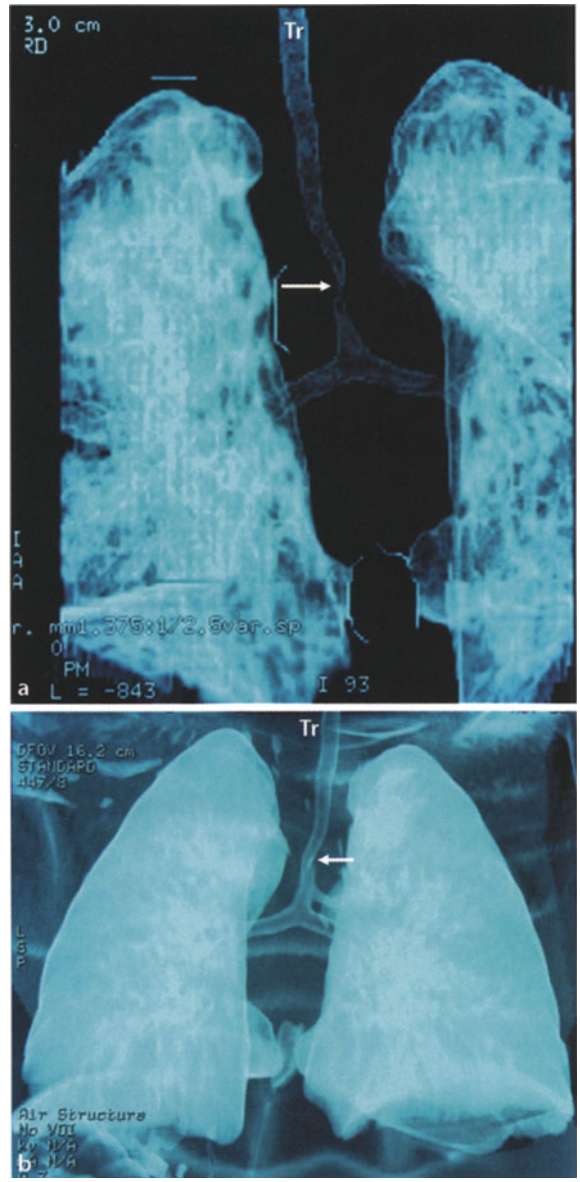

Fig. 2.15.3. Pulmonary artery sling: radiology. **a** Anteroposterior projection of the CT scan in the same patient an in Fig. 2.15.2 showing the associated long-segment tracheal stenosis with severe distal deviation and obstruction (white arrow), **b** anteroposterior projection of the magnetic resonance in the same patient showing the associated long-segment tracheal stenosis with severe distal deviation and obstruction (white arrow) (*Tr* trachea)

and morbidity are caused by the associated tracheomalacia and bronchomalacia as a consequence of the increasing duration of airway compression, early surgical treatment is indicated to reduce mortality and morbidity in these patients. Reimplantation of the

anomalous left pulmonary artery is the treatment of choice, with or without tracheal reconstruction according to degree, extension and duration of the airway involvement. In the presence of an associated malformation of the upper airway, its surgical treatment must absolutely be performed during the same operation for the vascular lesion. The airway surgery as well as the postoperative course are much more complicated if the surgical treatment of the airway has to be done during a re-do thoracotomy on a patient impossible to wean from the mechanical ventilation and tracheal intubation because of the presence of a nondiagnosed tracheal stenosis. Moreover, the surgical access to the trachea is optimal during the same cardiopulmonary by-pass used to treat the vascular malformation, allowing a shorter operation time for the airway reconstruction.

Surgical treatment

Division of the anomalous left pulmonary artery at the origin and its mobilization and subsequent reimplantation into the main pulmonary artery anterior to the trachea can be accomplished either with or without cardiopulmonary bypass, by means of a side-biting clamp applied to the side of the main pulmonary artery. The mobilization of the pulmonary artery is enhanced by the division of the ligamentum arteriosum and needs to be as much as possible extensive, in order to reduce tension on the subsequent anastomosis, therefore, reducing the risk for stenosis or occlusion of the reimplanted artery. The anomalous left pulmonary artery is separated at its origin from the right pulmonary artery, and the remaining opening in the right pulmonary artery is closed generally with direct suture; in very small children it might be necessary to use a small patch of autologous pericardium. Then the left pulmonary artery, well dissected and separated from the surrounding structures, including the trachea anteriorly and the esophagus posteriorly, is pulled into the left pleural space and then brought back into the pericardium through a window created in the pericardium posteriorly to the left phrenic nerve at the level of the main pulmonary artery. At this point the proximal end of the transected left pulmonary artery is end-to-side anastomosed to the left aspect of the main pulmonary artery, carefully avoiding twisting, kinking or traction.

The procedure of reimplantation of the anomalous left pulmonary artery has been reported via left or (very rarely) right thoracotomy, but the preferred approach is through a median sternotomy: this approach not only allows better mobilization of the pulmonary artery and the choice between the reimplantation with or without cardiopulmonary bypass, but also leaves the possibility for simultaneous tracheal reconstruction (of course with cardiopulmonary by-

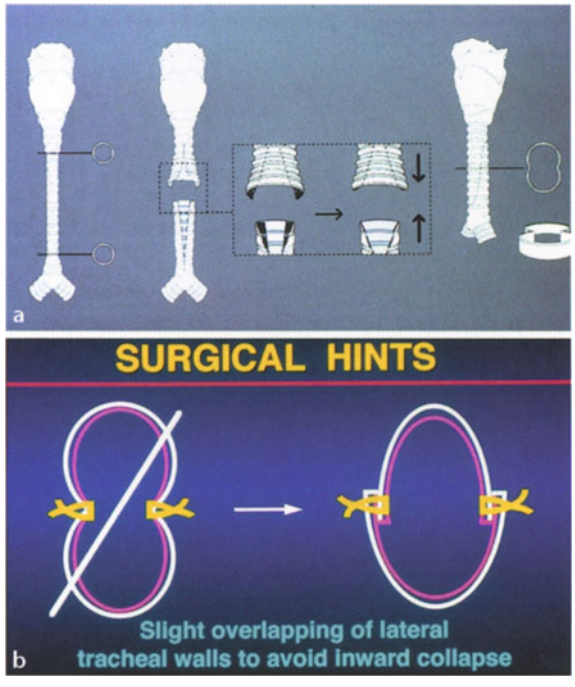

Fig. 2.15.4. Pulmonary artery sling: surgery. **a** Schematic drawing of the surgical technique of the slide tracheoplasty, **b** schematic drawing of the surgical technique of the slide tracheoplasty, showing overlapping of the lateral tracheal walls to avoid inward collapse (photographs courtesy of Dr. Philippe Monnier)

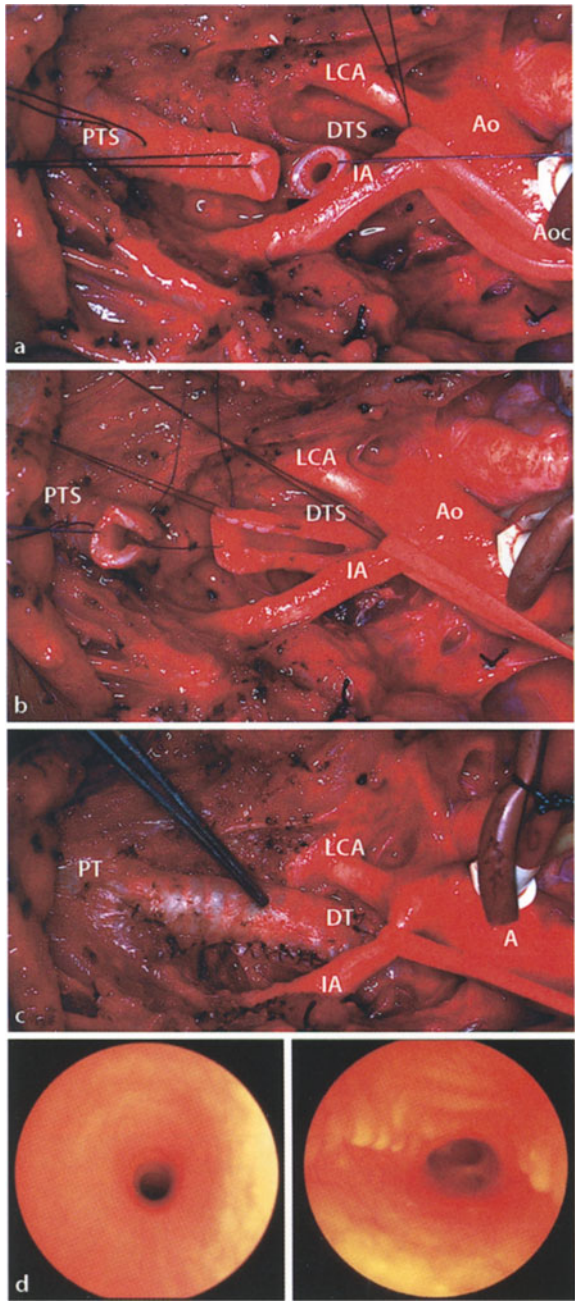

Fig. 2.15.5. Pulmonary artery sling: surgery. **a** Intraoperative photograph showing the surgical technique of the slide tracheoplasty; after extensive mobilization of the long segment tracheal stenosis and beginning of cardiopulmonary bypass, the trachea is divided in the middle of the long segment stenosis (*Ao* aorta, *Aoc* aortic cannula, *DTS* distal tracheal stump, *IA* innominate artery, *LCA* left carotid artery, *PTS* proximal tracheal stump), **b** longitudinal incision of the two stumps of the trachea over the entire longsegment stenosis, respectively on the posterior aspect of the proximal tracheal stump, and on the anterior aspect of the distal tracheal stump, **c** completed lateral anastomosis of the two stumps of the trachea over the entire length (*DT* distal trachea, *PT* proximal trachea), **d** tracheoscopy (photograph courtesy of Dr. Philippe Monnier)

pass) in the presence of complete tracheal rings with or without long-segment tracheal stenosis.

With regard to tracheal reconstruction, several techniques have been adopted for circular ring stenosis.

Resection is the procedure of choice for short stenoses (<1/3 of the tracheal length) without involvement of the carina or the main bronchi:

- For long segment stenosis, *pericardial patch tracheoplasty* was the gold standard for a long time, but now tends to be progressively replaced by slide tracheoplasty.
- *Slide tracheoplasty*, feasible even in infants in the presence of very long segment tracheal stenosis (Fig. 2.15.4, 2.15.5 and 2.15.6), has been demonstrated to provide the most reliable and consistent early and long-term results.
- Homograft tracheoplasty, prosthetic patch or costal cartilage tracheoplasty have been reported in a few cases with very inconsistent results.

Potential complications

Residual or recurrent stenosis (with also reported occlusion) at the origin of the reimplanted left pulmonary artery, residual or recurrent airway obstruction, tracheomalacia, recurrent respiratory infections.

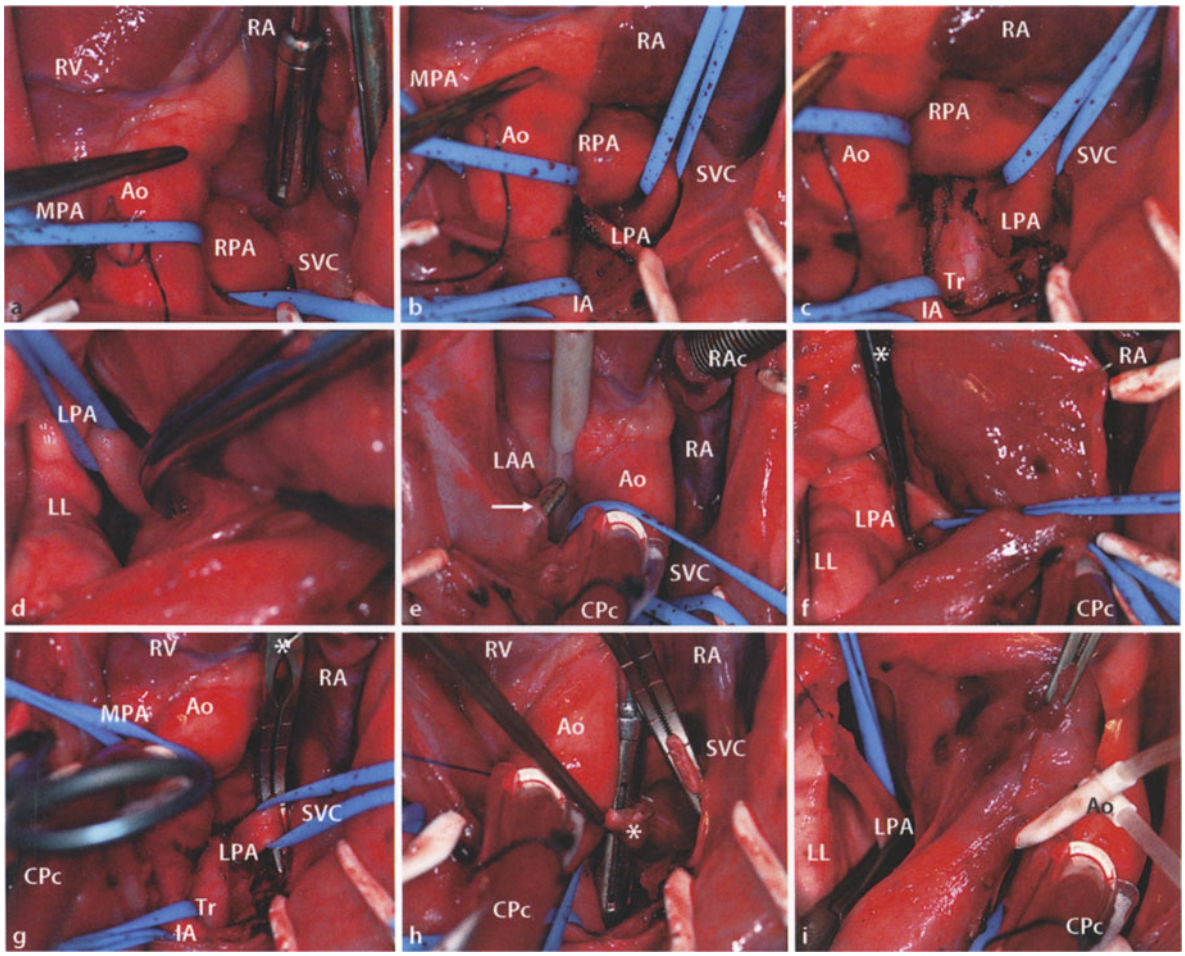

Fig. 2.15.6. Pulmonary artery sling: surgery. Intraoperative photograph of the same child as in Figs. 2.15.2 and 2.15.3 showing **a** the intrapericardial anatomy (*Ao* aorta, *MPA* main pulmonary artery, *RA* right atrium, *RPA* right pulmonary artery, *RV* right ventricle, *SVC* superior vena cava), **b** the anomalous left pulmonary artery arising from the right pulmonary artery, dissected free and controlled with a blue elastic vessel loop (*IA* innominate artery), **c** the progressive preparation of the anomalous left pulmonary artery arising from the right pulmonary artery, and its relationship with the anterior aspect of the trachea (*Tr* trachea), **d** the preparation of the distal intrapleural portion of the anomalous left pulmonary artery, dissected free and controlled with a blue elastic vessel loop (*LL* left lung), **e** after the cannulation for cardiopulmonary bypass, the preparation by a surgical instrument (white arrow) of a window created in the pericardium posteriorly to the left phrenic nerve at the level of the main pulmonary artery by an opening through the left pleura to create the new pathway for the left pulmonary artery, dissected free and controlled with a blue elastic vessel loop. (*CPc* cardioplegia cannula, *LAA* left auricular appendage, *RAc* right atrial cannula), **f** the distal occlusion of the left pulmonary artery, controlled by a blue elastic vessel loop, by a vascular clamp (white star), in the left pleural cavity, **g** the proximal occlusion of the left pulmonary artery, controlled by a blue elastic vessel loop, by a vascular clamp (white star), at its origin from the right pulmonary artery, **h** the division of the proximal left pulmonary artery from its origin from the right pulmonary artery; the distal stump (white star) is controlled with forceps, **i** the left pulmonary artery, well dissected and separated from the surrounding structures, including the trachea anteriorly and the esophagus posteriorly, pulled into the left pleural space.

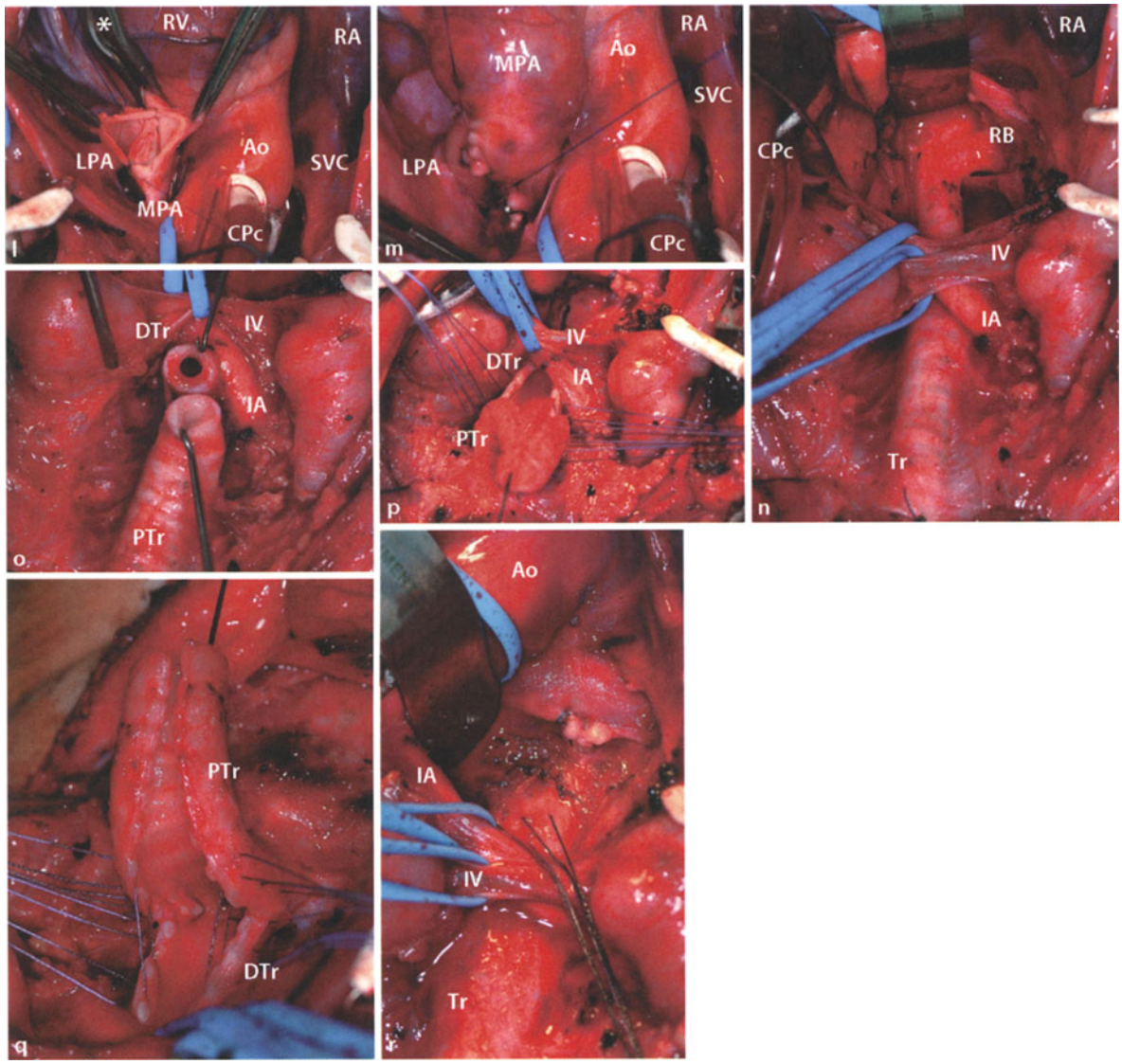

Fig. 2.15.6 l–r. **l** The posterior portion of the end-to-side anastomosis of the left pulmonary artery to a longitudinal incision performed on the right aspect of the main pulmonary artery, controlled with a vascular clamp (white asterisk), **m** the completed end-to-side anastomosis of the left pulmonary artery to the main pulmonary artery, **n** the preparation of the trachea with longsegment stenosis for the slide tracheoplasty; note the small size of the trachea in comparison with the normal size of the right bronchus; innominate artery and vein have been dissected free and controlled with blue elastic vessel loops (*IV* innominate vein, *RB* right bronchus), **o** the transversal division of the trachea at the middle of the long segment stenosis; note the very narrow internal lumen of the distal trachea (*DTr* distal trachea, *PTr* proximal trachea), **p** the first sutures to approximate the two stumps of the trachea after longitudinal opening of the anterior aspect of the distal stump and of the posterior aspect of the proximal stump, **q** the surgical details of the sutures approximating the two stumps of the trachea, showing the substantial increase of the tracheal size allowed by the slide tracheoplasty, **r** the completed slide tracheoplasty

Vascular ring

Incidence

Airway stenosis is present in 1–2% of children with congenital heart defects, and is more frequent in patients with Down syndrome (40%) than in the normal population. No gender prevalence has been detected.

Morphology

Several congenital vascular anomalies can determine airway compression at the level of the trachea or of the main bronchi (mostly the left), and esophageal compression, because of the presence of a *complete or incomplete vascular ring* constituted by the aortic arch and its branches. Generally a vascular ring is due to the presence of a double aortic arch or to the presence of a right aortic arch, but a right aortic arch may occur without forming a vascular ring; the presence or absence of a vascular ring in the setting of a right aortic arch depends upon the branching of the brachiocephalic vessels and the location of the ductus arteriosus.

Frequent forms:

- *Double aortic arch:* double aortic arch is one of the two most common forms (45–65% of cases) of complete vascular rings, with trachea and esophagus completely encircled by connected segments of the aortic arch and its branches; as the name implies, this anomaly consists of two aortic arches, an anterior and leftward arch and a posterior and rightward arch.
  Various forms of double aortic arch exist: both arches may be patent, or an atretic (but persistent) segment may exist at one of several locations in either arch. Generally the descending aorta is left sided although it may be right sided or in the midline. The right arch, generally dominant (in 50–75% of patients), gives origin to the right common carotid and right subclavian arteries either as an innominate artery or as two separate vessels. The left arch, which gives origin to the left common carotid and left subclavian arteries, in the majority of patients is patent, but it may be hypoplastic or atretic beyond the origin of either the left common carotid (rarely) or the left subclavian artery (more frequently). In the latter case a fibrous cord joins the descending aorta, where it emerges from behind the esophagus to become the left descending aorta near the insertion of the ligamentum arteriosum. Note that the right recurrent laryngeal nerve must pass around the right aortic arch, rather than being in its usual location around the right subclavian artery. In 15–25% of patients the left arch is dominant, and in these cases the right arch is almost always patent, while in 15–25% of patients right and left aortic arch present with almost equal size.
- *Right aortic arch with anomalous origin of the left subclavian artery and left ductus arteriosus or ligamentum arteriosum:* a right aortic arch that gives origin, in sequence, to the left common carotid, the right common carotid, the right subclavian, and the left subclavian arteries. The left subclavian artery, last branch originating from the aortic arch, passes behind the esophagus and then gives origin to the ductus arteriosus or ligamentum arteriosum, which passes anteriorly to connect to the proximal left pulmonary artery, thereby completing the vascular ring.
- *Right aortic arch with mirror-image branching and left retroesophageal ductus arteriosus or ligamentum arteriosum:* a right aortic arch that gives origin, in sequence, to the left innominate artery (left common carotid with left subclavian), the right common carotid, and the right subclavian artery. The final branch, often arising from a prominent ductus diverticulum, is a patent ductus arteriosus or ligamentum arteriosum that passes leftward, behind the esophagus, and then anteriorly

to connect the left pulmonary artery. When there is mirror image branching, if the ligamentum arteriosum arises from the innominate artery to pass to the origin of the left pulmonary artery, this does not result in a complete vascular ring. Bilateral ductus arteriosus has also been reported.

- *Anomalous innominate artery:* the innominate artery originates more posterior than usual from the left aortic arch and crosses posteriorly the trachea, causing compression of the anterior tracheal wall (*innominate artery compression syndrome*).
- *Pulmonary artery sling* (see above).

Rare forms

- *Right or left retroesophageal aortic arch.*
- *Right aortic arch with anomalous left subclavian artery with or without aortic coarctation*: this combination is technically not a complete vascular ring, but it may cause symptoms similar to a ring because of the presence of a right-sided patent ductus arteriosus or ligamentum arteriosum, contributing to the formation of an incomplete vascular ring.
- *Situs inversus with left aortic arch and right ligamentum arteriosum.*
- *Dominant left aortic arch, mirror-image branching, right descending aorta, and atretic right aortic arch:* a dominant left aortic arch is extremely rare; with this rare form of vascular ring, the arch vessels arise normally from the normal-sized left aortic arch, while the right arch is atretic.
- *Left aortic arch, right descending aorta, and right-sided ligamentum arteriosum to right pulmonary artery:* the reported branching sequence from the left aortic arch is the right common carotid, left common carotid, left subclavian, and, finally, right subclavian as a fourth branch from the proximal descending aorta.

Associated anomalies

A vascular ring is generally an isolated cardiac malformation, with ventricular septal defect and tetralogy of Fallot probably the most common associated anomalies. Very rarely it can be associated with anomalous left subclavian artery, left or right patent ductus arteriosus, aortic coarctation, univentricular heart, pulmonary atresia with ventricular septal defect (unusual), double outlet right ventricle, truncus arteriosus, transposition of the great arteries.

Since the major clinical impact of the presence of a vascular ring in children is tracheal compression, altered tracheal geometry has been demonstrated in all symptomatic children with vascular rings, with smaller dimensions (area, shorter and longest diameters) compared with asymptomatic children.

Esophageal compression can be associated with the airway compression, while esophageal atresia is sometimes found in association with double aortic arch.

A vascular ring can be associated (in approximately 20–25% of patients) with deletion of chromosome 22q11.2 or DiGeorge syndrome (CATCH-22 syndrome = cardiac defect, abnormal face, thymic hypoplasia, cleft palate, hypocalcemia, microdeletion of band 22q11), as well as occasionally with VATER syndrome (= vertebral defects, anal atresia, tracheoesophageal fistula with esophageal atresia, renal and radial anomalies) and CHARGE syndrome (= coloboma, heart disease, atresia choanae, retarded growth, retarded development and/or central nervous system anomalies, genital hypoplasia, ear anomalies and/or deafness).

Pathophysiology

Vascular rings, in contrast to a pulmonary artery sling, encircle both the esophagus and the trachea and, therefore, may result in obstruction of both. Nevertheless, the mere presence of a ring does not guarantee that

there will be compression, since the extent of respiratory impairment depends on the severity of airway compression and degree of tracheomalacia, which can vary considerably; rarely rings may remain asymptomatic for life and not require any intervention.

Airway compression is more frequently associated with double aortic arch than with other forms of vascular ring, and the compression of the trachea and/or one of the main bronchi is often more severe. Tracheomalacia and bronchomalacia are frequently associated as a consequence of the airway compression, particularly in the presence of Down syndrome, malnutrition, long segment tracheal stenosis with complete cartilaginous tracheal rings (see chapter "Pulmonary artery sling"), increasing duration of the vascular compression with recurrent respiratory symptoms or assisted mechanical ventilation.

Airway obstructions in children with Down syndrome are complicated by upper airway obstructive pathologies such as nasopharyngeal, oropharyngeal and subglottic disease.

Esophageal compression is generally progressive and provides symptoms typical of dysphagia.

Diagnosis

Clinical pattern: presentation of symptoms depends on the severity of airway and/or esophageal compression and the presence of associated anomalies; nonpositional inspiratory stridor, noisy breathing, expiratory wheezing, "seal barky" cough, respiratory distress, recurrent lower or upper respiratory tract infections and apnea are due to airway compression; progressive dysphagia with increasing difficulty for solid food, with swallowing difficulties, choking, reflux and recurrent vomiting are related to esophageal compression; failure to thrive is the generalized consequence of airway and/or esophageal compression; young patients may experience episodes often referred to as "apparent life-threatening events" or "death spells", with an acute apneic or severe obstructive crisis accompanied by cyanosis.

Electrocardiogram: not diagnostic; usually it is normal, with the exception of abnormalities due to the presence of associated cardiac anomalies.

Chest X-ray: the presence of a right aortic arch is already suggestive of an associated vascular ring; the lateral projection shows tracheal bowing, as well as tracheal narrowing at the level of the aortic arch; bilateral tracheal indentations may be appreciated on overpenetrated radiograms; vascular rings produce a posterior indentation of the esophagus evident on barium swallow on the lateral projection, and bilateral indentations in anteroposterior projection, with indentations at different levels and of a different size: the more superior and usually larger indentation given by the right arch and the more inferior and usually smaller indentation given by the left arch; it is important to distinguish a double arch from the retroesophageal subclavian or ligamentum arteriosum, based on the angulation of the esophageal impression (Fig. 2.15.7); video esography simultaneously with angiography allows the visualization of the pulsatile cause of the obstruction.

Echocardiogram: echocardiography, with suprasternal, high parasternal and subcostal views, generally allows accurate definition of the anatomy of a vascular ring, particularly when imaging is combined with Doppler color flow.

Cardiac catheterization: nowadays aortography is rarely needed for diagnosis of vascular rings, and is indicated in the presence of associated cardiac malformations; with or without simultaneous tracheobronchoscopy, it allows the identification of the anomalous vascular structures and the relationship with the airways (Fig. 2.15.8).

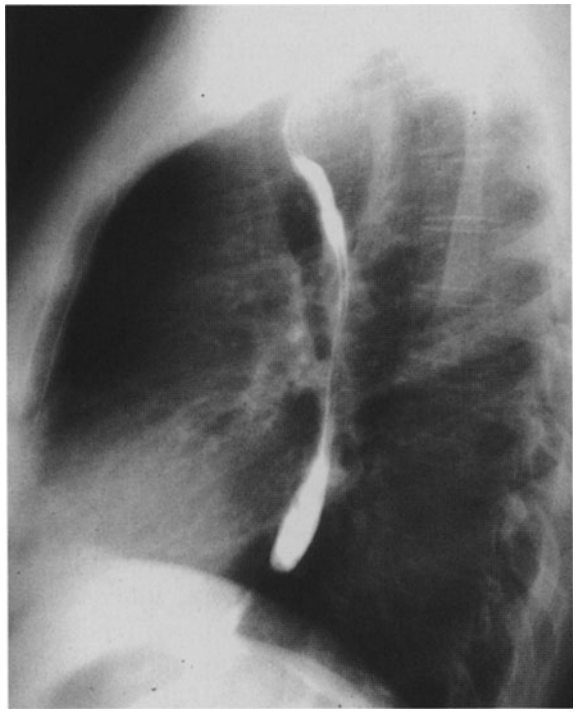

Fig. 2.15.7. Vascular ring: radiography. Barium swallow esophagogram, showing the esophageal indentation due to the simultaneous presence of right aortic arch and anomalous left subclavian artery

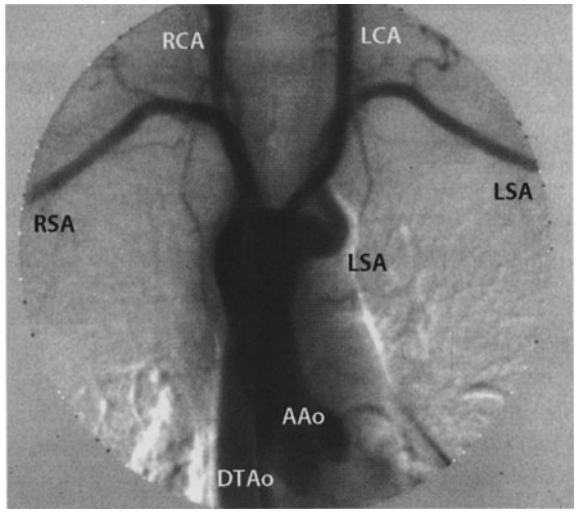

Fig. 2.15.8. Vascular ring: angiography. Contrast injection in the ascending aorta of the same patient as in Fig. 2.15.7 showing the simultaneous presence of right aortic arch and anomalous left subclavian artery (*AAo* ascending aorta, *DTAo* descending thoracic aorta, *LCA* left carotid artery, *LSA* left subclavian artery, *RCA* right carotid artery, *RSA* right subclavian artery)

Bronchoesophagoscopy: is the gold diagnostic standard in the diagnosis of an external compression of the upper airway and digestive tract by a vascular structure; the endoscopic location of a pulsatile extrinsic compression of the trachea and/or the esophagus allows the identification of the type of vascular malformation; showing pulsatile compression of the posterior and lateral tracheal walls (anterior and lateral tracheal walls in the presence of right aortic arch).

Computed tomography and magnetic resonance imaging: also very useful in order to define the anatomical details of both the airway and the vascular structures.

Indications for surgical treatment

Since the untreated severe respiratory obstructions occurring in the first few months of life are generally fatal before the age of 1 year, the general agreement is that if either respiratory or dysphagic symptoms are present, surgical division of the ring is indicated. Only if the child is asymptomatic may surgery be deferred. Preoperatively the child should be given maximal nutritional support as well as general respiratory care, including chest physiotherapy and appropriate treatment of respiratory infection. Surgery should not be unduly delayed because of the presence of a respiratory infection, as division of the ring, which allows more adequate clearing of respiratory secretions, is the most effective treatment of infection. Furthermore, delayed surgery may result in either sudden death of further tracheobronchial damage.

In the presence of a vascular ring, the most convenient surgical approach is through a left posterolateral thoracotomy, because the majority (more than 95% of the cases) of vascular rings consist of a dominant right arch. This approach can be modified by the presence of a dominant left aortic arch, extremely rare, and of associated congenital heart defects. Right thoracotomy is used only in the presence of innominate

artery compression syndrome. If a double aortic arch is present, it is important to be aware preoperatively which of the arches is dominant: in the vast majority of cases is the right aortic arch.

Surgical treatment (without cardiopulmonary bypass)

For the division of the vascular ring, the chest is entered through the fourth intercostal space, the left lung is retracted anteriorly, and the mediastinal pleura is reflected in the area of the left arch and ligamentum arteriosum or ductus arteriosus. After dissection, identification and control of all the vessels with elastic vessel loops, the segment to be divided is controlled with vascular clamps (Fig. 2.15.9). After division, the stumps of the vessel are oversewn with continuous sutures. If the segment to be divided is clearly atretic, division between double ligatures can be sufficient. After division the two stumps generally retract briskly, indicating the tension with which the ring has been surrounding the esophagus and trachea. In all cases the ligamentum arteriosum or the patent ductus arteriosus must also be divided. Frequently, there are additional fibrous strands passing across the esophagus and/or the trachea, and all these have to be divided to completely relieve the compression (Figs. 2.15.10 and 2.15.11). In the rare case requiring approach through a right thoracotomy, the same principles are applied. The right recurrent laryngeal nerve will pass around the right-sided ligamentum arteriosum and should be carefully visualized and preserved. Techniques of video-assisted thoracoscopic division have recently been developed as an alternative approach to the division of vascular ring.

In the case of persistent compression of trachea and/or main bronchus by the aorta or a pulmonary artery branch, "arteriopexy" performed by suturing the retroesophageal aortic segment to the prevertebral fascia or through the sternum; "extension" of the aor-

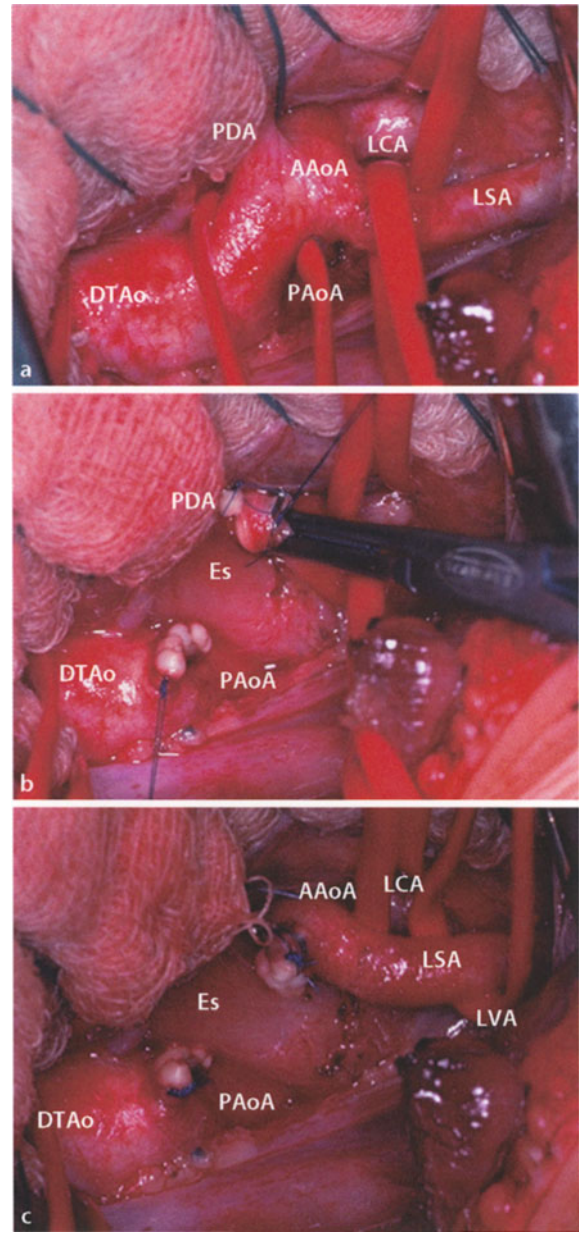

Fig. 2.15.9. Vascular ring: surgery. Intraoperative photograph in an infant with double aortic arch and patent ductus arteriosus; **a** surgical approach through left posterlateral thoracotomy in the fourth intercostal space; left lung retracted anteriorly with the white sponge (*AAoA* anterior aortic arch, *DTAo* descending thoracic aorta, *LCA* left carotid artery, *LSA* left subclavian artery, *PAoA* posterior aortic arch, *PDA* patent ductus arteriosus), **b** patent ductus arteriosus and anterior aortic arch have been divided and the stumps oversewn; the divided proximal stump of the anterior aortic arch is still controlled with a vascular clamp; the esophagus has been largely decompressed by the division of the anterior aortic arch, as demonstrated by the distance of the two stumps (*ES* esophagus), **c** final appearance after division of the patent ductus arteriosus and of the anterior aortic arch, with the esophagus now completely free (*LVA* left vertebral artery)

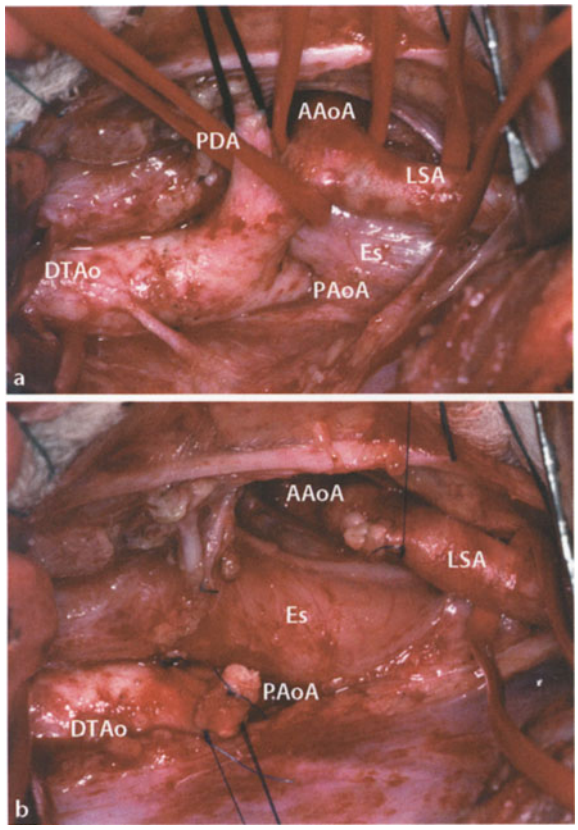

Fig. 2.15.10. Vascular ring: surgery. **a**, **b** Intraoperative photograph in a child with double aortic arch and patent ductus arteriosus, same surgical approach as for the patient as in Fig. 2.15.9 (*AaoA* anterior aortic arch, *DTAo* descending thoracic aorta, *ES* esophagus, *LSA* left subclavian artery, *PAoA* posterior aortic arch, *PDA* patent ductus arteriosus)

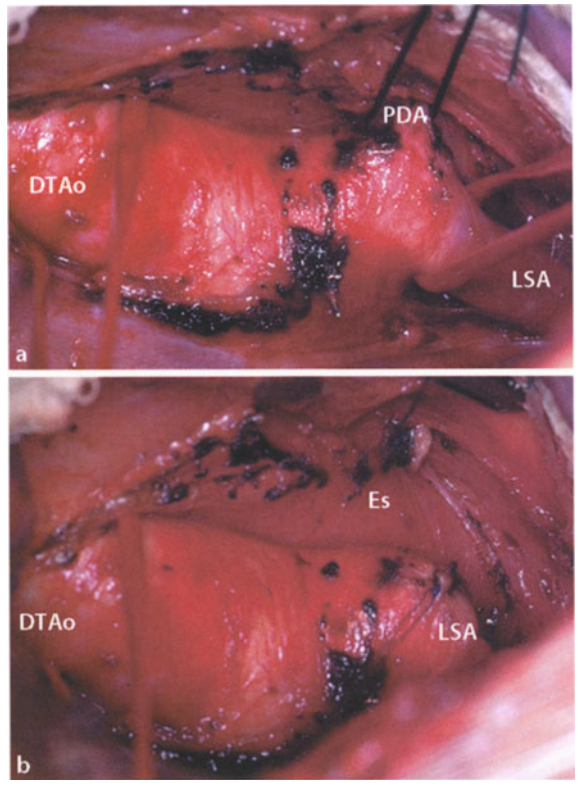

Fig. 2.15.11. Vascular ring: surgery. **a** Intraoperative photograph in a young adult patient with Kommerel diverticulum and patent ductus arteriosus, **b** the patent ductus arteriosus has been divided and the esophagus is free (*DTAo* descending thoracic aorta, *LSA* left subclavian artery, *Es* esophagus, *PDA* patent ductus arteriosus)

ta or the pulmonary artery by a tubular prosthesis may be necessary.

In the presence of *innominate artery compression syndrome*, the approach is through right anterior thoracotomy with suspension of the innominate artery to the posterior aspect of the sternum; an alternative technique is the transection of the innominate artery at the origin and its reimplantation on the aorta in a more proximal position.

In the presence of tracheal or bronchomalacia, tracheal suspension or bronchial suspension can be accomplished with traction and fixation to the chest wall: when these procedure are performed with simultaneous intraoperative tracheobronchoscopy, immediate control of the obtained relief of airways compression is accomplished. If the suspension procedure does not provide satisfactory relief, other procedures may be required, like resection if localized pathology, reinforcement of tracheal wall by implantation of rib cartilage or other allogenic resorbable material.

In the presence of associated intracardiac anomalies requiring simultaneous repair with cardiopulmonary bypass through median sternotomy, the division of the vascular ring is performed during the same procedure from a frontal approach, also advisable in the presence of associated tracheal or esophageal lesions requiring simultaneous surgical treatment.

Potential complications

Since in the young infant with severe respiratory symptoms there is likely to be an element of tracheo/bronchomalacia associated with the long-standing compression by the ring during in utero development, it should be anticipated that all respiratory symptoms will not be immediately relieved, even after complete relief of the external airway compression; in fact it may be several months before the child is free of respiratory symptoms.

Nevertheless, residual or recurrent airway obstruction is possible, as well as recurrent respiratory infections, chylothorax (lesion to the thoracic duct), diaphragmatic paresis/paralysis (injury to the phrenic nerve) or vocal cord paresis/paralysis (injury to the recurrent laryngeal nerve).

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Chapter 2.16 Cardiac tumors

Incidence

Primary cardiac tumors are very rare clinical entities, with benign neoplasms occurring three to four times more often (70–90%) than malignant tumors (10–30%). Only series of myxomas in adults have been reported, while very few series involving different types of primary cardiac tumors in children have been reported. In this chapter only types of pediatric cardiac tumors occurring in the pediatric age will be considered. Metastatic cardiac neoplasms are not included. The ability to detect cardiac tumors with noninvasive diagnostic procedures even during the prenatal period has led to an apparent increase in this diagnosis, with incidences reported from 0.06% cases to 0.32%, while the reported autopsy series was limited to 0.027–0.08%.

Morphology (Fig. 2.16.1)

Benign tumors: *rhabdomyoma* is the most common primary cardiac tumor in the pediatric age, particularly in the neonatal period; 75% of cases are diagnosed in infancy. Rhabdomyomas, a benign tumor of cardiac myocytes, typically occur as multiple tumors (in 90% of the patients), particularly when associated with tuberous sclerosis, and rarely as a sporadic lesion. They also present as circumscribed lesions, with a yellowish-gray color, frequently with the multiple lesions in both ventricles and in the interventricular septum, but can be found also in the atria, at the cavoatrial junction and on the epicardial surface. They vary from small (few millimeters) to extremely large (several centimeters). Despite not being noncapsulated, they generally do not embolize.

Fibroma is the second most common cardiac tumor in the pediatric age, with at least 40% diagnosed in infancy; it has been called also fibromatosis or fibroelastic hamartoma. This tumor, usually solitary and large (multiple ventricular fibromas have been also reported), circumscribed, firm, gray-white, frequently centrally calcified, often involves the left ventricular free wall or septum, while right ventricular involvement is more rare. Sometimes the fibroma incorporates proximal segments of coronary arteries, precluding complete surgical removal, particularly when located at the superior portion of the interventricular septum and the crux of the heart posteriorly.

Papillary fibroelastoma is the third most common primary heart tumor in the pediatric age. It is generally a relatively small lesion, but it may involve structures like a mitral or aortic valve leaflet; since papillary fibroelastoma is usually a pedunculated and mobile mass, there is a frequent incidence of systemic embolism, sometimes responsible for sudden death.

Myxoma is the most frequent cardiac tumor in the adult population, with an incidence of 3/1000 patients, while in the pediatric population it occurs less frequently, accounting for only 9–15% of all cardiac tumors diagnosed from birth to adolescence. Myxomas appear more frequently in females. The cardiac myxoma is generally solitary, polypoid and pedunculated, friable, gelatinous, but with a broad base. The most frequent lo-

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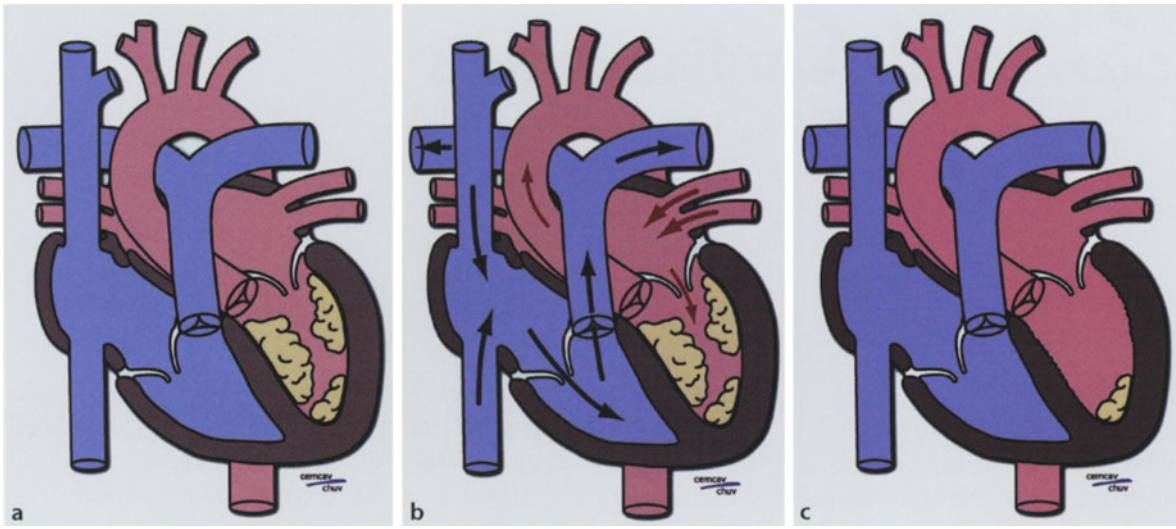

Fig. 2.16.1. Cardiac tumors: **a** morphology, **b** pathophysiology, **c** surgery

calization of myxomas is in the left atrium (75% of cases) where most of the time arises from the atrial septum (at the limbus of the fossa ovalis), followed by the right atrium (20%), and very rarely in the ventricles or on cardiac valves. These types of tumor present with a recurrence rate much higher (21%) than sporadic myxomas (1%).

Teratoma is single, encapsulated, and it appears most often in the pericardium; in neonates (50% of teratomas are diagnosed in the neonatal period) the teratoma can be larger than the patient's own heart and determine cardiac tamponade or pulmonary compression; there is potential for malignant transformation.

Hemangioma is a benign proliferation of endothelial cells, also known as vascular tumor and can occur in any part of the heart.

Other types of tumors have been less frequently reported in children and include lipoma, mesothelioma, leiomyoma, pheochromocytoma, Purkinje cell tumor and benign cystic tumors.

Malignant tumors: in the pediatric age these are all extremely rare. *Sarcomas* are the most frequent primary cardiac malignant neoplasms, with various types of sarcoma; the angiosarcoma is the most common histologic type (most frequently in males), with reported cases of rhabdomyosarcoma, leiomyosarcoma, liposarcoma, fibrosarcoma, osteosarcoma, mixosarcoma. Rhabdomyosarcoma grows invasively, metastatizes and can recur; these tumors are more common in children because they may arise from embryonic cell remains in the septum. Malignant fibrous histiocytoma, lymphoma and leiomyosarcoma have also been reported.

All these malignant tumors seed blood directly, therefore metastases are common and widespread, found in as many as 88% of patients with malignant cardiac tumors.

Associated anomalies

Cardiac rhabdomyomas are associated very frequently (30 to 86% of patients) with tuberous sclerosis, and very rarely with congenital heart defects. Conversely, a rhabdomyoma is diagnosed in 50–60% of patients with tuberous sclerosis. Ventricular septal defect, parachute mitral valve, Ebstein's anomaly, pulmonary atresia, and patent ductus arteriosus have been reported in association with primary cardiac tumors.

Pathophysiology

Large-sized tumors can obstruct the inflow and/or the outflow ventricular tracts, whereas small lesions can involve the conducting system causing arrhythmias. Rhabdomyomas generally do not grow, and partial or complete spontaneous regression of the mass of the tumors have been documented in at least 50% of the patients. Fibroma has a very slow but continuous growth, and may cause conduction disturbances and/or obstruction to the left ventricular outflow tract. Because of the gelatinous consistency of myxomas, thrombosis and tumor fragments are prone to embolize; since the left atrium is the most frequent location of cardiac myxomas, systemic embolization is the most frequent clinical complication.

Diagnosis

Clinical pattern: the symptoms are generalized, nonspecific, and depend on number, size and location of the tumor(s), and may vary between abnormal heart murmur, arrhythmias, hydrops, dyspnea, syncope, easy fatigability, angina, low cardiac output, cyanosis (due to right-to-left shunt because of right ventricular inflow or outflow tract obstruction associated with patent foramen ovale) and severe heart failure; sudden death before surgical removal has been reported in pediatric patients; systemic embolization can be the first sign of the presence of a cardiac myxoma (where peripheral embolization is reported to occur in as many as 45–70% of patients) or papillary fibroelastoma (with sudden death).

Electrocardiogram: in children with ventricular rhabdomyoma ventricular hypertrophy is due to the electrically active tissue of the tumor, even in absence of ventricular pressure overload or ventricular dilatation; ST-T changes may be induced by coronary arteries compression by the tumor(s); all types of arrhythmias have been reported in the presence of cardiac tumor, accordingly with the location; in most cases electrocardiographic abnormalities tend to disappear with the regression of the tumors.

Chest X-ray: cardiomegaly may be present in cases with hemodynamic obstruction; distortion of the cardiac silhouette can result from any type of cardiac tumor; certain neoplasm, like fibroma, can contain calcium, detected on plain chest X-ray.

Echocardiogram: echocardiography is the most useful and expedient diagnostic tool, with precise recognition of location, extent and characteristics (single or multiple, intracavitary or intramuscular, solid or cystic) of the tumor, appearing as filling defect(s) in the ventricular cavity (Fig. 2.16.2), and its hemodynamic consequences (by Doppler evaluation); prenatal diagnosis is also diagnostic with increasing frequency; the diagnosis of cardiac rhabdomyoma is made primarily by echocardiographic evidence of multiple cardiac tumors, with associated tuberous sclerosis.

Cardiac catheterization: not required to made the diagnosis; endomyocardial biopsy is seldom performed, because of the high risk of metastasis during the procedure in the presence of a malignant tumor; it can be useful to show the coronary arteries in the presence of extrinsic compression.

Indications for surgical treatment

Benign tumors: although the behavior of cardiac tumors is unpredictable, these forms are generally resectable; indication for surgical treatment is given in the presence of severe obstruction to the right or left ventricular inflow and/or outflow (Figs. 2.16.3–2.16.5), in the presence of arrhythmias or of systemic embolization. This is true particularly for rhabdomyomas, since regression of the size of the tumors with growth has been frequently reported for multiple cardiac rhabdomyomas. The younger the age at diagnosis, the higher the chances for spontaneous regression are, complete regression being more

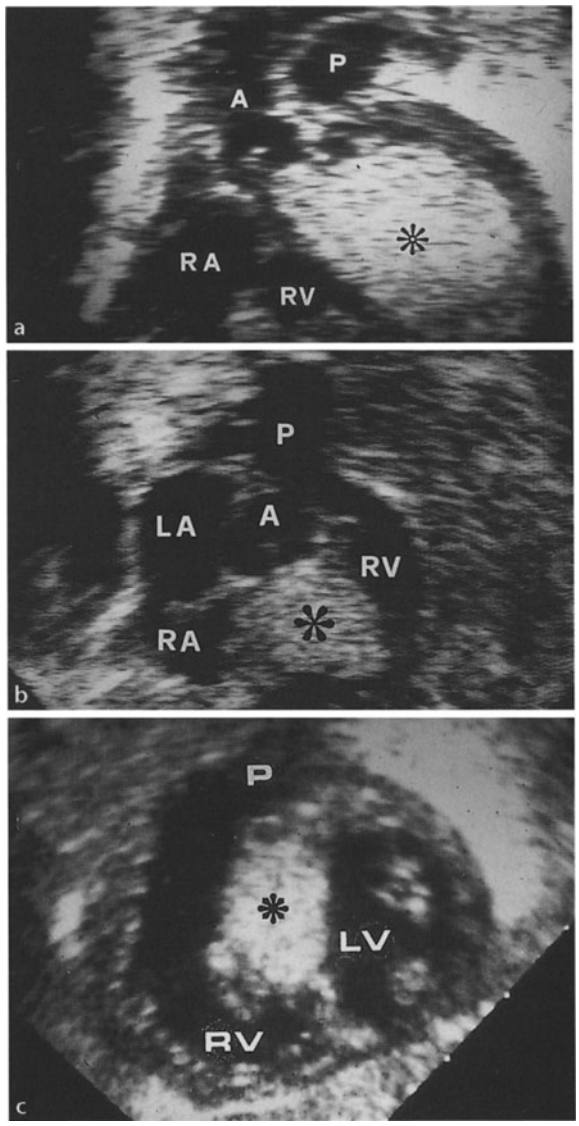

Fig. 2.16.2. Cardiac tumor: echocardiography. a Long-axis subxiphoid view showing a very large tumor (asterisk) originating from the interventricular septum and occupying almost the entire left ventricular cavity (*A* aorta, *P* pulmonary artery, *RA* right atrium, *RV* right ventricle) (reproduced with permission from Marino B, Thiene G (1990) Atlante di anatomia ecocardiografica delle cardiopatie congenite, USES, Firenze), **b** right oblique subxiphoid view showing a very large tumor (asterisk) obstructing the right ventricular cavity (*A* aorta) (photograph courtesy of Dr. Bruno Marino), **c** left oblique subxiphoid view showing a very large tumor (asterisk) located on the interventricular septum (*LV* left ventricle) (reproduced with permission from Marino B, Thiene G (1990) Atlante di anatomia ecocardiografica delle cardiopatie congenite, USES, Firenze)

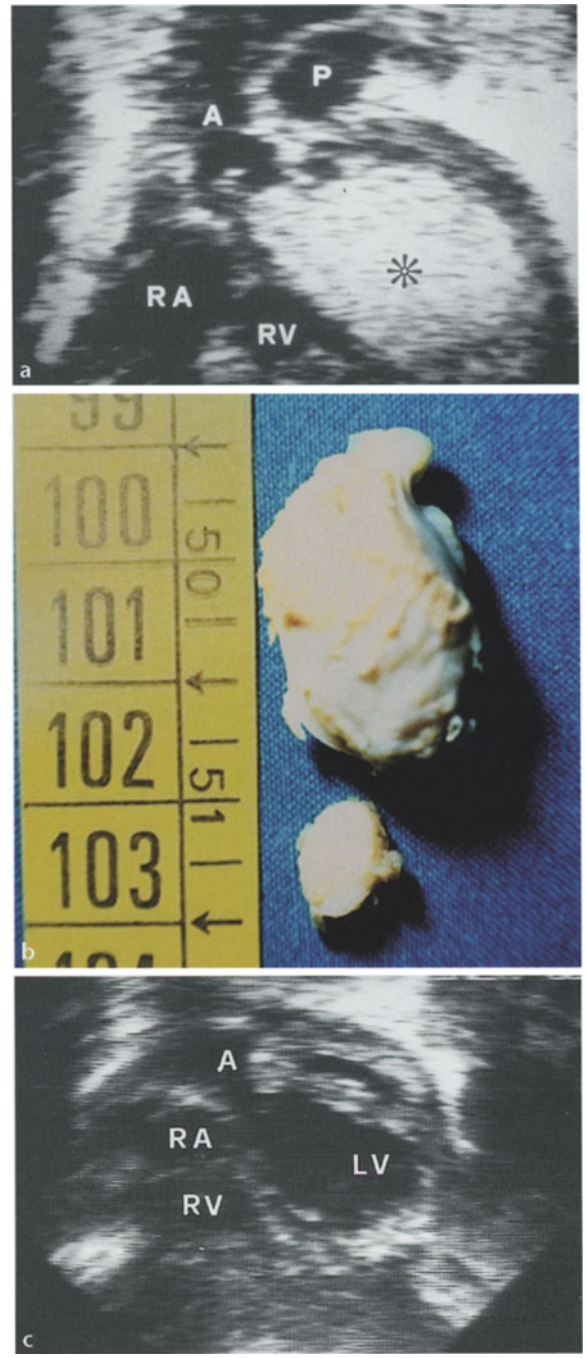

Fig. 2.16.3. Cardiac tumor: surgery. **a** Preoperative echocardiographic long-axis subxiphoid view in a neonate showing a very large tumor (asterisk) occupying almost the entire left ventricular cavity (*A* aorta, *P* pulmonary artery, *RA* right atrium, *RV* right ventricle), **b** Intraoperative photograph of the same neonate as in **a** showing the macroscopic appearance of the tumors after surgical excision, **c** postoperative echocardiography in the same neonate as in **a** showing the left ventricular cavity completely free (*LV* left ventricle) (**a–c** reproduced with permission from Corno AF, de Simone G, Catena G, Marcelletti C (1984) Cardiac rhabdomyoma: surgical treatment in the neonate. J Thorac Cardiovasc Surg 87:725–731)

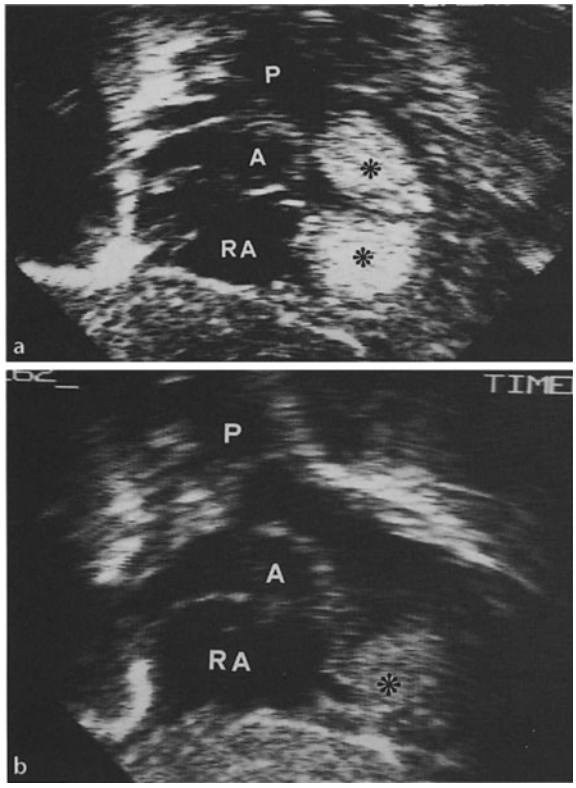

Fig. 2.16.4. Cardiac tumor: surgery. **a** Preoperative echocardiographic right oblique subxiphoid view in another neonate showing two very large tumors (asterisks) occupying almost the entire right ventricular cavity, **b** Postoperative echocardiographic right oblique subxiphoid view in the same neonate as in **a** showing the right ventricular outflow tract completely free, and a residual mass (asterisk) attached to the tricuspid valve (*A* aorta, *P* pulmonary artery, *RA* right atrium) (reproduced with permission from Corno AF, de Simone G, Catena G, Marcelletti C (1984) Cardiac rhabdomyoma: surgical treatment in the neonate. J Thorac Cardiovasc Surg 87:725–731)

frequent within the first 4 years of life. In these patients the prognosis depends upon the presence of associated tuberous sclerosis.

Malign tumors: cardiac sarcomas are extremely aggressive, and without treatment the survival is less than one year. Unfortunately these forms are seldom resectable, and only tumor debulking may be possible because of the extent of the local spread and invasion, or because of the frequent distal metastases. Heart transplant, with radiation and chemotherapy, has been considered in some of these patients with unresectable cardiac masses.

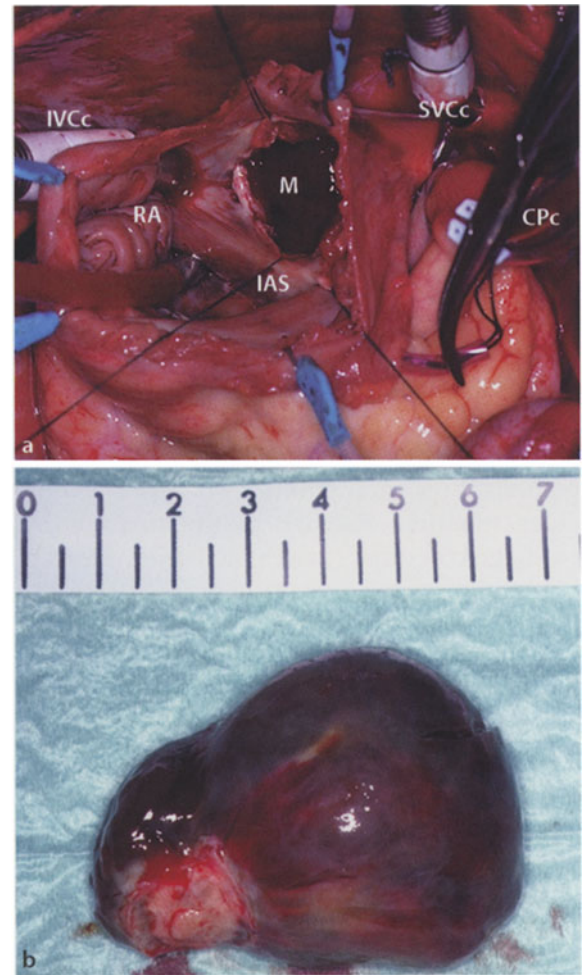

Fig. 2.16.5. Cardiac tumor: surgery. **a** Intraoperative photograph showing the transseptal approach to remove a left atrial myxoma (*CPc* cardioplegia cannula, *IAS* interatrial septum, *IVCc* inferior vena cava cannula, *M* myxoma, *RA* right atrium, *SVCc* superior vena cava cannula), **b** intraoperative photograph showing the entirely resected left atrial myxoma, with the white fibrous tissue corresponding to the insertion on the interatrial septum

Surgical treatment (on cardiopulmonary bypass)

The surgical approach and technique of resection entirely depend upon the size and location of the tumor(s); whenever possible the resection is performed through a right or left atriotomy, or from the aorta or the pulmonary artery. Right and/or left ventriculotomy may be required to resect large-sized or multiple tumors obstructing the ventricu-

lar inflow and/or outflow. The surgical excision, particularly in neonates and infants, is limited to the area of the tumor, because extensive myocardial involvement of vital structures (valves, conduction tissue, coronary arteries) can preclude complete excision of the tumor. Occasionally intrapericardial tumors can be excised without cardiopulmonary bypass.

Potential complications

Incomplete resection, systemic or pulmonary embolization, arrhythmias, reduced ventricular function, damage to the surrounding cardiac structures (valves, atrial or ventricular septum, conduction tissue), recurrence.

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Chapter 2.17 Aortico-left ventricular tunnel

Incidence

The true incidence is unknown, although 2 cases among 1754 (=0.1%) patients with congenital heart defects have been reported in a 10 year period. Male to female ratio is 2:1.

Morphology (Figs 2.17.1 and 2.17.2)

Aortico-left ventricular tunnel is an extremely rare congenital malformation, consisting of an abnormal paravalvular communication between an abnormal aneurysmal dilatation to the aortic root and the upper portion of a sinus of Valsalva, above the sino-tubular junction, and the left ventricle, bypassing the aortic valve. The tunnel can originate from the right coronary sinus (most frequently) or from the left coronary sinus (rarely).

Extremely rare is the occurrence of an aorto-right ventricular tunnel. Size and shape of the aortico-ventricular tunnel are extremely variable.

The following classification has been proposed to categorize aortic-left ventricular tunnel:

- *Type I:* simple tunnel with a slitlike opening at the aortic end, without aortic valve distortion.
- *Type II:* large extracardiac aortic wall aneurysm of the tunnel with an oval opening at the aortic end, with or without aortic valve distortion.
- *Type III:* intracardiac aneurysm of the septal portion of the tunnel, with or without right ventricular outflow tract obstruction.
- *Type IV:* combination of *types II* and *III*.

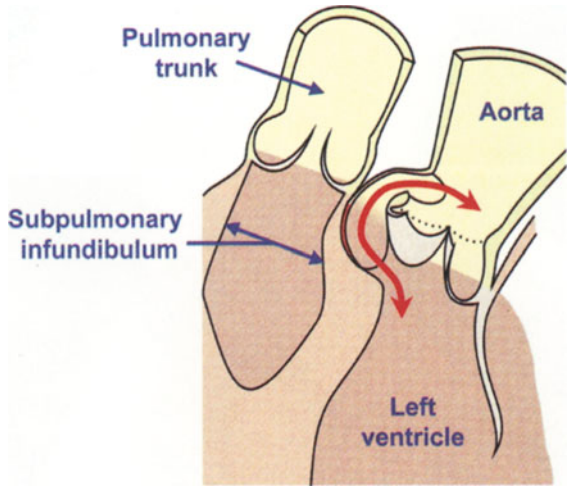

Fig. 2.17.1. Aortico-left ventricular tunnel: morphology. Diagram showing the essential anatomy of a tunnel bypassing the hinge of the aortic valve to produce an aortic-left ventricular tunnel (reproduced with permission from: McKay R, Anderson RH, Cook AC (2002) Cardiol Young 12:563–580)

Associated anomalies

Because of the relationship between the aortico-ventricular tunnel and the origin of the coronary arteries, relatively frequent are the associations with coronary artery malformations: atresia of a coronary artery orifice, anomalous origin of the left or right coronary artery. Other associated cardiac anomalies are bicuspid aortic stenosis, aortic atresia, atrial septal defect, ventricular septal defect, pulmonary valve stenosis, aneurysm of sinus of Valsalva, patent ductus arteriosus.

A. F. Corno, *Congenital Heart Defects*

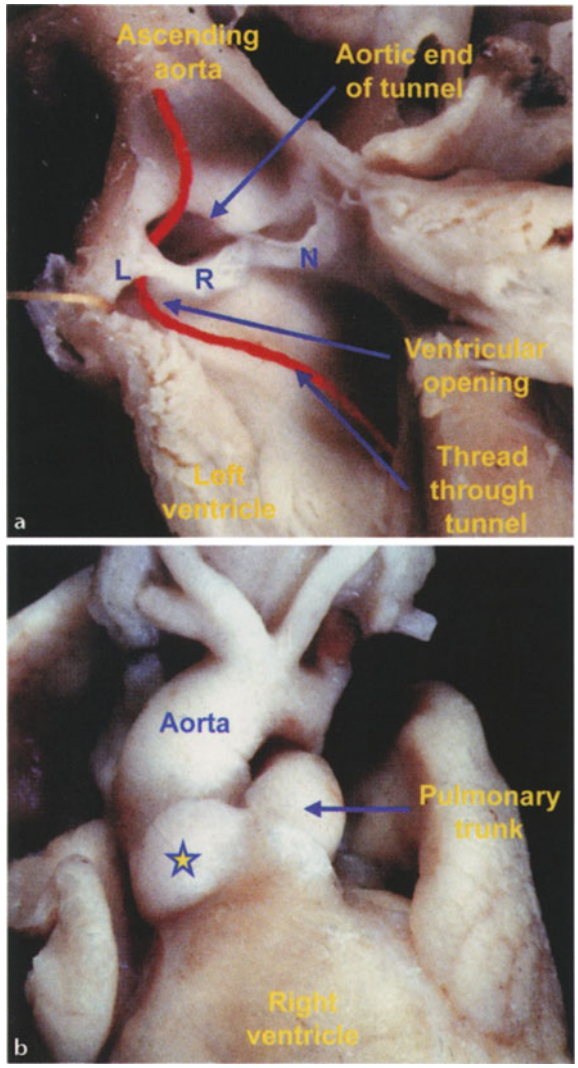

Fig. 2.17.2. Aortico-left ventricular tunnel: morphology. **a** The tunnel, extending from above the right coronary aortic sinus to open within the fibrous triangle between the left and right coronary leaflets of the aortic valve, runs within the tissue plane that separates the sinuses of the aortic valve from the free-standing muscular sub-pulmonary infundibulum (*L* left coronary aortic leaflet, *N* non-coronary aortic leaflet, *R* right coronary aortic leaflet), **b** the heart shown is the same as illustrated in **a**. Note the bulge made by the tunnel (starred) between the aorta and the pulmonary trunk (reproduced with permission from: McKay R, Anderson RH, Cook AC (2002) Cardiol Young 12:563–580)

Pathophysiology

The pathophysiological pattern depends upon the cross sectional area of the tunnel and the degree of the often associated aortic valve regurgitation. Hemodynamics equivalent to a severe aortic regurgication is the typical pattern, with subsequent aneurysmal dilatation and progressive left ventricular failure. Because the tunnel traverses the upper portion of the interventricular septum, there is the potential for an aneurysmal tunnel to create an obstruction to the right ventricular outflow tract. Involvement of the left coronary artery, because of the position of the tunnel, has been reported, but only very rarely.

Diagnosis

Clinical pattern: age at presentation and severity of symptoms depend upon the size of the tunnel, and are extremely variable, from asymptomatic adults (rarely), to severely symptomatic neonates and infants because of congestive heart failure or sudden death. The clinical signs are the same as for aortic regurgitation, with left ventricular dysfunction: loud systolic and diastolic thrill and murmur, with preserved second heart sound; wide systemic pulse pressure with bounding peripheral pulses.

Electrocardiogram: left-axis deviation, left ventricular hypertrophy, possible T-wave inversion, ST changes in the presence of myocardial ischemia.

Chest X-ray: cardiomegaly, broad upper mediastinal shadow from the dilated ascending aorta; the esophagogram shows the typical anterior indentation of the esophagus.

Echocardiogram: parasternal long-axis view shows a ventricular septal dropout immediately below the aortic valve, with tubular communication between the aorta and the left ventricle bypassing the aortic valve, and with massive regurgitation through the tunnel in systole demonstrated by Doppler color flow; short-axis views demonstrate the presence of aortic-ventricular tunnel opening into the right ventricle.

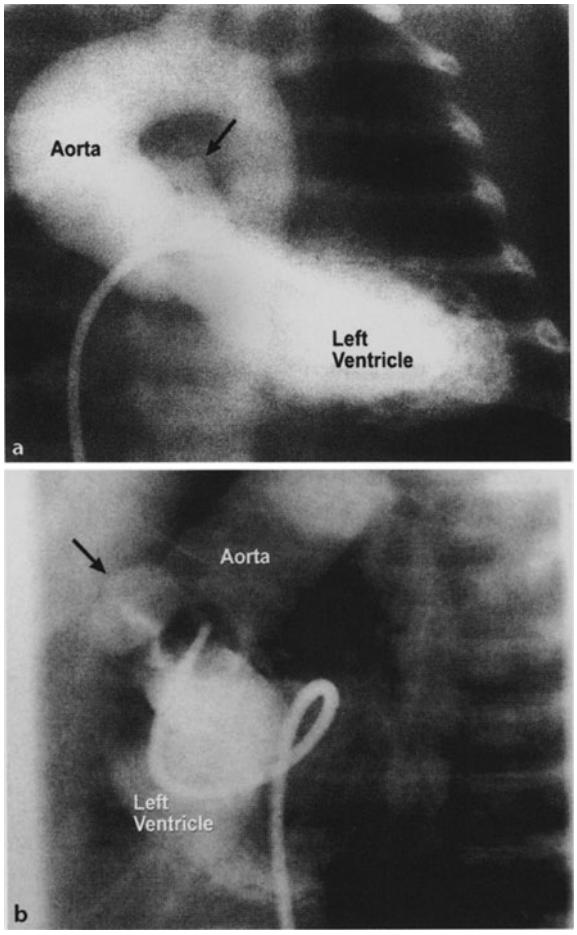

Fig. 2.17.3. Aortico-left ventricular tunnel: angiography. **a** Left ventriculogram in the antero-posterior projection and **b** in the lateral projection, showing a tunnel (arrow) extending from the aorta to the left ventricle in a neonate (reproduced with permission McKay R, Anderson RH, Cook AC (2002) Cardiol Young 12:563–580)

Cardiac catheterization: performed only to rule out uncommon associated cardiac anomalies or to obtain precise anatomical details (Fig. 2.17.3).

Indications for surgical treatment

In an infant with an hemodynamically important tunnel, the natural history is characterized by rapidly progressive congestive heart failure and sudden death within the first few months of life.

The surgical treatment is therefore indicated as early as possible, in order to prevent damage to the aortic valve with progression of the aortic regurgitation, and left ventricular dilatation with heart failure. The most important factor leading to a major aortic valve regurgitation after surgical repair is an inherent weakness at the junction between the membranous part of the ventricular septum and the anterior part of the aortic annulus. This weakness, either structural or secondary to the turbulence created by the to-and-fro movement of blood through the tunnel, will change the geometry of the aortic annulus, especially at the right coronary sinus area. With the growth of the aortic root and even after surgical repair, the weakness can result in the dysfunction and prolapse of the right aortic cusp.

The goals of surgery are to eliminate the ventricular volume overload without damaging the aortic valve, the coronary arteries and the conduction tissue, and to relieve any left or right ventricular outflow tract obstruction.

Surgical treatment (on cardiopulmonary bypass)

The surgical treatment is on cardiopulmonary bypass, and it is very important to prevent and avoid left ventricular distension because of rapid aortic run-off. The surgical technique consists, after exposure of the defect through a transverse aortotomy and careful identification of the origin of the coronary arteries, in the direct or patch (PTFE, Teflon, pericardium) closure of the aortic side only or both ends of the tunnel (Fig. 2.17.4). Patch closure of both ends of the tunnel is the preferred technique to prevent distortion of the sinus, and subsequent progression of the aortic valve regurgitation, a potential consequence of direct closure, and recurrencies, presenting with higher incidence after closure of the aortic end only. Associated aortic valve replacement has been reported in the presence of severe lesion of the aortic valve, not suitable to reparative procedure.

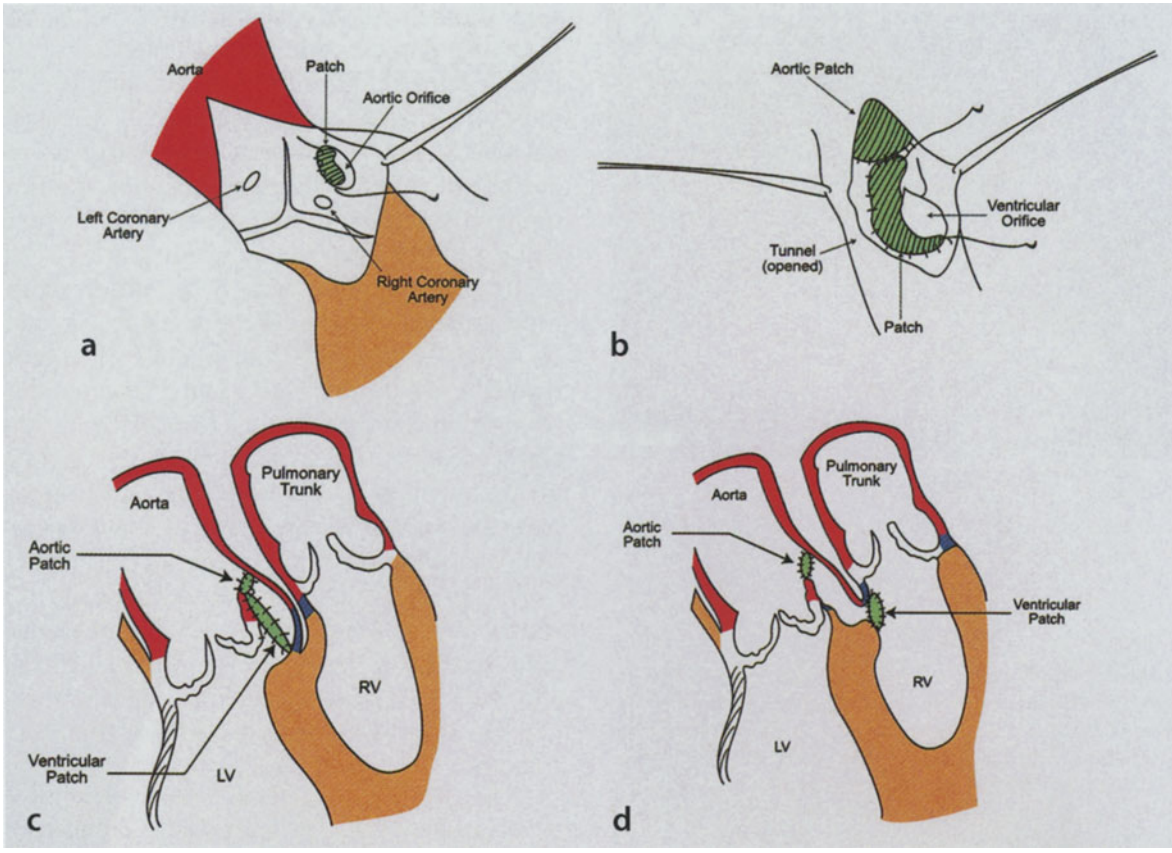

Fig. 2.17.4. Aortico-left ventricular tunnel: surgery. The diagrams illustrate the most commonly employed technique for repair of an uncomplicated tunnel running from the aorta to the left ventricle (**a**, **b**, **c**) or the right ventricle (**a**, **d**). The aortic orifice (**a**) is closed through an aortotomy by suturing a patch to the sinotubular ridge and the aortic wall, having identified the orifices of both coronary arteries. The tunnel itself is opened vertically (**b**). The ventricular orifice is closed using a second patch, which is sutured to the ventricular myocardium, and, in the case of tunnels ending within the left ventricle, also to the fibrous wall of the unsupported aortic sinus, and the bottom of the first patch. The walls of the tunnel are then approximated over the patches, and the aortotomy is closed. Figure **c** shows the completed repair of a tunnel to the left ventricle, with the aortic leaflet now supported by the patches. Figure **d** shows a completed repair for a tunnel terminating in the right ventricle (reproduced with permission from McKay R, Anderson RH, Cook AC (2002) Cardiol Young 12:563–580)

Potential complications

Residual or recurrent blood flow through the tunnel, residual or progressive aortic valve regurgitation (generally correlated with the age and the type of repair, but also with the presence or development of annulo-aortic ectasia), obstruction to the coronary artery perfusion, complete atrio-ventricular block.

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Subject index